PAEDIATRICS

Genetic and Metabolic Disorders — Questions

1. **For a normal husband and wife, the first child was diagnosed to be having cystic fibrosis. What is the likely percentage of chance for the second child to be affected with cystic fibrosis?**

   A. 0 %
   B. 25 %
   C. 50 %
   D. 100 %

2. **The following condition is seen in which among the following condition?**

   A. Partial trisomy 18
   B. Partial trisomy 13
   C. Partial trisomy 21
   D. Partial trisomy 22

3. **An infant presents with failure to thrive, past history of seizures and presently having skin rash. Investigations show metabolic acidosis, increased blood ketone levels. This child is likely to be suffering from?**

   A. Propionic acidemia
   B. Urea cycle disorder
   C. PKU
   D. Multiple carboxylase deficiency

4. **Inheritance of congenital adrenal hyperplasia is?**

   A. Autosomal dominant
   B. Autosomal recessive
   C. X-linked recessive
   D. Sporadic condition

5. **Mousy odour of urine is seen in?**

   A. Isovaleric acidemia
   B. Maple system urine disease
   C. Tyrosinemia
   D. Phenylketonuria

6. **All are true about Fragile X syndrome except?**

   A. Large head
   B. Large nose
   C. Large ear
   D. Large testis

7. **A 16-year old female with Down syndrome is brought to the physician for behavioural changes and has developed urinary incontinence. On examination, she is hypotonic, hyper-reflexic with a positive Babinski sign. She is ataxic. What is the likely co-morbidity suspected in this child?**

   A. Alzheimer disease
   B. Embolic stroke
   C. Atlanto-axial instability
   D. Spinal cord infarction

8. **Mechanism not involved in Down syndrome genetics?**

   A. Robertsonian translocation
   B. Genomic imprinting
   C. Non-disjunction of maternal chromosomes
   D. Mosaicism

9. **Most common cause of intestinal obstruction in Down syndrome is?**

   A. Duodenal atresia
   B. Jejunal atresia
   C. Hirschsprung disease
   D. Colonic atresia

10. **Not seen in Edward syndrome?**

    A. Association with advanced maternal age
    B. Digit abnormalities
    C. Life expectancy upto 5 years in >10%
    D. Rocker bottom feet

11. **A baby at birth was found to have microcephaly, cleft palate, hypotelorism and scalp defects. What is the likely diagnosis?**

    A. Trisomy 21
    B. Trisomy 18
    C. Trisomy 13
    D. Trisomy 8

12. **The following condition at birth is most commonly seen in which among the following condition?**

    A. Noonan syndrome
    B. Turner syndrome
    C. Fragile X syndrome
    D. Down syndrome

13. **A male child with webbed neck, short stature, antimongoloid slant of eyes and obesity will have which among the following underlying genetic abnormality?**

    A. 45, XO genotype
    B. Trisomy 11
    C. 45, XO/46, XY mosaicism
    D. Somatic gene mutation

14. **Trident hand with champagne glass pelvis is seen in?**

    A. Cleidocranial dysostosis
    B. Achondroplasia
    C. Hypophosphatasia
    D. Osteogenesis imperfecta

15. **Most commonly involved gene in Rett syndrome is?**

    A. RET
    B. P53
    C. RB
    D. MECP-2

1. b	2. d	3. d	4. b	5. d	6. b	7. c	8. b	9. a	10. c
11. c	12. b	13. d	14. b	15. d					

16. Tyrosinemics are more susceptible to develop?

A. Adenocarcinoma colon
B. Melanoma
C. Hepatocellular carcinoma
D. Retinoblastoma

17. A 7-month-old baby with hyponatremia, emesis, hepatosplenomegaly, failure to thrive and CT shows adrenal gland calcification?

A. Wolman's disease
B. Testosterone
C. Adrenal hemorrhage
D. Pheochromocytoma

18. Treatment of multiple carboxylase deficiency is?

A. Pyridoxine
B. Biotin
C. Thiamine
D. Folic acid

19. Urine becomes dark on standing in which condition?

A. Alkaptonuria
B. Cystinuria
C. Fabry's disease
D. Tyrosinemia

20. Globoid cells are a diagnostic feature of?

A. Krabbe's disease
B. Progressive multifocal leukoencephalopathy (PMFL)
C. Tay-sach's disease
D. Metachromatic leukodystrophy (MLD)

21. Nitisinone is useful in the management of?

A. Phenylketonuria
B. Alcaptonuria
C. Homocystinuria
D. Tyrosinemia

22. A baby that was apparently normal at birth begins to show a delay in motor development by 3 months of age. At one year of age, the child begins to develop spasticity and writhing movements. At age three, compulsive biting of fingers and lips and head-banging appear. At puberty, the child develops arthritis, and death from renal failure occurs soon after. This patient's condition is due to an enzyme deficiency in which of the following biochemical pathways?

A. Ganglioside metabolism
B. Tyrosine metabolism
C. Purine metabolism
D. Pyrimidine metabolism

23. A newborn vomits after each feeding of milk-based formula, and does not gain weight. Biochemical testing reveals a severe deficiency of galactose-1-phosphate uridyl transferase, consistent with homozygosity. If this condition goes untreated, what is the likely outcome for this patient?

A. Benign disease except for cataract formation
B. Chronic emphysema appearing in early adulthood
C. Chronic renal failure appearing in adolescence
D. Death in infancy

24. Dried blood spot (DBS) test is used in neonates to check for?

A. Inborn errors of metabolism
B. Arterial blood gas analysis
C. Blood group
D. Creatinine and bilirubin

25. Enzyme replacement therapy is available for?

A. Albinism
B. Gaucher disease
C. Niemann-pick disease
D. Metachromatic leukodystrophy

16. c	17. a	18. b	19. a	20. a	21. d	22. c	23. d	24. a	25. b

1. **B. 25 %**

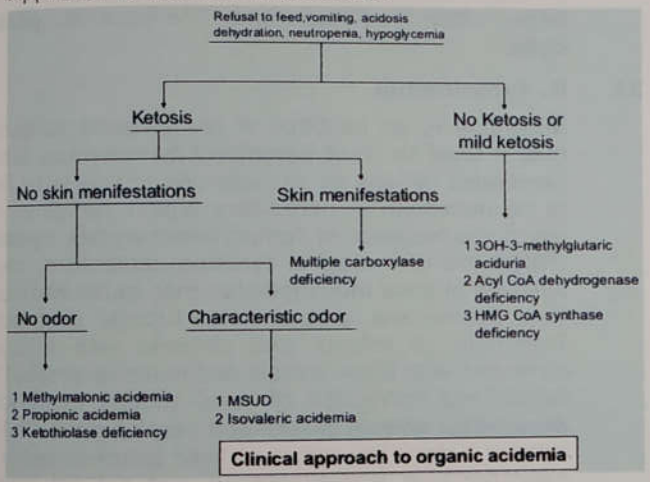
Cystic fibrosis is an autosomal recessive disorder. Now, since both parents are unaffected, the chance of having an affected offspring is 25% in each pregnancy (Not overall)!!! So chance will be 25% in each pregnancy in this case.

2. **D. Partial trisomy 22**

The patient is showing coloboma of iris, seen in cat eye syndrome. Most patients harbor a small supernumerary bisatellited marker chromosome (sSMC) that results in partial tetrasomy or partial trisomy in chromosome 22. The 3 main visible characteristics of this condition are preauricular anomalies, anal atresia, and iris coloboma

3. **D. Multiple carboxylase deficiency**

Approach to acidosis with ketosis



4. **B. Autosomal recessive**

According to Nelson 20th Ed, Congenital adrenal hyperplasia (CAH) is a family of autosomal recessive disorders of cortisol biosynthesis, with 21-hydroxylase deficiency being seen in >90% cases.

5. **D. Phenylketonuria**

Explanation:

Odours attributed to IEMs	
Phenylketonuria (PKU)	Musty, mousy
Tyrosinemia	Musty, Cabbage like
Maple syrup urine disease	Sweet, Maple syrup
Isovaleric acidemia	Sweaty feet
Multiple carboxylase deficiency	Cat urine

6. **B. Large nose**

Fragile sites are regions of chromosomes that show a tendency for separation, breakage, or attenuation under particular growth conditions. One fragile site that has clinical significance is the one on the distal long arm of chromosome Xq27.3 associated with the fragile X syndrome. Fragile X accounts for 3% of males with intellectual disability The main clinical manifestations of fragile X syndrome in affected males are intellectual disability, autistic behavior, macro-orchidism, i.e. **large testes**, hyperextensible finger joints, and characteristic facial features, including **large head, large ears** and broad jaw.

7. **C. Atlanto-axial instability**

Up to 15% of children with Down syndrome have misalignment of cervical vertebra C1, which places them at risk for spinal cord injury with neck hyperextension or extreme flexion due to atlanto-axial instability. Any damage or cervical spine injury can produce UMN signs with atxia, as seen in the question above.

8. **B. Genomic imprinting**

Genetics of Down syndrome

- Maternal chromosomal non-disjunction during meiosis: 95%
- Translocations: 4%
- Mosaicism: 1%

9. **A. Duodenal atresia**

Duodenal atresia is MC GIT manifestation in Down syndrome, manifesting as neonatal intestinal obstruction.

10. **C. Life expectancy upto 5 years in >10%**

Life expectancy is below 6 months in Edward syndrome and always less than 1-2 years. Other features can be seen in Edward syndrome. For details, refer to class notes or see chapter 1 of DAMS-CRS.

TRISOMY 18
Small and premature appearance
Tight palpebral fissures
Narrow nose and hypoplastic nasal alae
Narrow bifrontal diameter
Prominent occiput
Micrognathia
Cleft lip or palate
Microcephaly
Congenital heart disease (e.g., VSD, PDA, ASD) Short sternum, small nipples
Limited hio abduction
Clinodactyly and overlapoing fingers; index over 3rd, 5th over 4th; closed fist
Rocker-bottom feet
Hypoplastic nails
Severe developmental delays and prenatal and postnatal growth restriction
Premature birth, polyhydramnios
Inguinal or abdominal hernias
Only 5% live >1 yr

11. **C. Trisomy 13**

These features are characteristic of Patau syndrome, i.e. Trisomy 13.

TRISOMY 13
HEAD AND FACE
Scalp defects (e.g., cutis aplasia)
Microphthalmia, corneal abnormalities
Cleft lip and palate in 60%-80% of cases
Microcephaly
Microphthalmia
Sloping forehead
Holoprosencephaly (arhinencephaly)
Capillary hemangiomas
Deafness
CHEST
Congenital heart disease (e.g., VSD, PDA, and ASD) in 80% of cases
Thin posterior ribs (missing ribs)

12. B. Turner syndrome

This is a child with dorsal lymphedema at birth, classically seen in Turner syndrome.

13. D. Somatic gene mutation

This is a child with Noonan syndrome, which is due to somatic gene mutation in PTPN-11 and RAS genes.

14. B. Achondroplasia

A trident hand is a description where the hands are short with stubby fingers, with a separation between the middle and ring fingers. It can be seen in various chondrodysplasias including achondroplasia. The **champagne glass pelvis** is a helpful sign in achondroplasia in which the iliac blades are flattened, giving rise to a **pelvic** inlet that resembles a **champagne glass**. It is a feature of achondroplasia.

15. D. MECP-2

Rett syndrome is caused by mutations in MeCP2, a transcription factor that binds to methylated CpG islands and silences transcription.

16. C. Hepatocellular carcinoma

In tyrosinemia, an acute hepatic crisis commonly heralds the onset of the disease and is usually precipitated by an intercurrent illness that produces a catabolic state. Most hepatic crises resolve spontaneously but may progress to liver failure and death. Between the crises, varying degrees of failure to thrive, hepatomegaly, and coagulation abnormalities often persist. Cirrhosis and eventually hepatocellular carcinoma occur with increasing age.

17. A. Wolman's disease

Wolman Disease (also known as Wolman's Disease, early onset LAL Deficiency, and Lysosomal acid lipase deficiency is a rare genetic disorder caused by a deficiency of an enzyme known as lysosomal acid lipase (LAL or LIPA). Clinical features become apparent in the first weeks of life and include failure to thrive, relentless vomiting, abdominal distention, steatorrhea, and hepatosplenomegaly. **Calcification of the adrenal glands** occurs in about 50% of patients. Death usually occurs within the first 6 months of life.

18. B. Biotin

Multiple carboxylase deficiency is caused by defects in the incorporation of the water-soluble B-complex vitamin biotin into or its release from four apocarboxylases acting on 3-methylcrotonyl-CoA, propionyl-CoA, acetyl-CoA, and pyruvate. Large doses of biotin (10–30 mg/day) usually produce rapid clinical improvement and almost complete disappearance of abnormal urine organic acids.

19. A. Alkaptonuria

Alkaptonuria (AKU) is a rare disorder of autosomal recessive inheritance. It is caused by a mutation in a gene that results in the accumulation of homogentisic acid (HGA). Characteristically, the excess HGA means sufferers pass dark urine, which upon standing turns black.

20. A. Krabbe's disease

In Krabbe's disease, some galactosylceramide in the form of filamentous or tubular inclusions accumulates in brain macrophages which are transformed into large, often multinucleated, PAS-positive, **globoid cells**.

21. D. Tyrosinemia

Nitisinone is an inhibitor of the tyrosine catabolism that is used to treat hereditary tyrosinemia, type 1. Developed initially as an herbicide, it was later found to be beneficial in hereditary type 1 tyrosinemia in which the absence of fumarylacetoacetate hydrolase (FAH), the fifth step in tyrosine catabolism, causes build-up of toxic intermediates that cause early onset liver failure and severe renal tubular dysfunction. Treatment of infants and children with nitisinone combined with a low protein diet led to reversal of liver failure and correction of renal tubular dysfunction, allowing for normal growth and development. Children treated before the onset of liver failure avoided the need for liver transplantation and did not develop hepatocellular carcinoma, common outcomes before the availability of this agent.

22. C. Purine metabolism

The patient is a classic case of Lesch-Nyhan syndrome, an X-linked disorder due to severe deficiency of the purine salvage enzyme hypoxanthine-guanine phosphoribosyl transferase (HPRT). This defect is associated with excessive de novo purine synthesis, hyperuricemia, and the clinical signs and symptoms described in the question stem. The biochemical basis of the often-striking self-mutilation behaviour (which may require restraints and even tooth extraction) has never been established. Treatment with allopurinol inhibits xanthine oxidase and reduces gouty arthritis, urate stone formation, and urate nephropathy. It does not, however, modify the neurologic/ psychiatric presentation.

23. D. Death in infancy

Galactosemia occurs in two very different clinical forms. Deficiency of galactokinase produces very mild disease with the only significant complication being cataract formation. In contrast, homozygous deficiency of galactose-1-phosphate uridyl transferase produces severe disease culminating in death in in-

fancy. In addition to galactosemia and galactosuria, these patients have impaired renal tubular resorption leading to aminoaciduria, gastrointestinal symptoms, hepatosplenomegaly, cataracts, bleeding diathesis, hypoglycemia, and mental retardation. Pathologically, the CNS shows neuronal loss and gliosis and the liver shows fatty change progressing to cirrhosis. Benign disease with cataract formation (choice A) is characteristic of galactokinase deficiency. Chronic emphysema (choice B) is not associated with homozygous galactose-1-phosphate uridyltransferase deficiency, but rather with alpha 1-antitrypsin deficiency. Impaired tubular reabsorption (producing aminoaciduria) is seen within a few days or weeks of feeding milk to an infant with severe galactosemia, as opposed to chronic renal failure appearing in adolescence (choice C).

24. A. Inborn errors of metabolism

For screening of a neonate, the baby's blood is collected in five circles on the newborn screening card. This is called a heel stick or dried blood spot (also called DBS) sample. The DBS is used to test for a specific group of conditions. Small pieces of the DBS are punched out and used to perform the newborn screen.

25. B. Gaucher disease

Enzyme replacement therapy (ERT) for type 1 Gaucher disease includes imiglucerase (Cerezyme), velaglucerase alfa (VPRIV), and taliglucerase alfa (Elelyso).

26. All are associated with malignancy except?
A. Down syndrome
B. Fragile X syndrome
C. Bloom syndrome
D. Fanconi's anemia

27. Most common benign tumour in children is?
A. Hemangioma
B. Teratoma
C. Leukemia
D. Osteoma

28. Most common malignancy in children is?
A. Wilm's tumour
B. Lymphomas
C. ALL
D. Brain tumours

29. A 1 yr old boy presenting with mass in abdomen and bone pain with raccoon eyes. What is the diagnosis?
A. Neuroblastoma
B. Wilm's tumor
C. Rhabdomyosarcoma
D. Nephroblastoma tumor

30. Which among the following is the most common inherited malignancy in children?
A. Infantile leukemia
B. Neuroblastoma
C. Retinoblastoma
D. Brain tumour

31. Suprasellar calcification is seen in
A. Craniopharyngioma
B. Medulloblastoma
C. Papilloma of the choroid plexus
D. Oligodendroglioma

32. Most common posterior fossa tumor in children is:
A. Medulloblastoma
B. Glioblastoma multiforme
C. Astrocytoma
D. Meningioma

33. While bathing her 2-year old child, a mother feels a mass in his abdomen. A thorough medical evaluation of the child reveals aniridia, hypospadias, horse shoe kidney and hemihypertrophy. The most likely diagnosis for the child is?
A. Neuroblastoma
B. Wilms tumour
C. Hepatoblastoma
D. Rhabdomyosarcoma

34. Most important prognostic factor for wilms' tumour is?
A. Stage
B. Size
C. Histology
D. Genetic abnormality

35. Most common malignant brain tumour in children is?
A. Glioblastoma
B. Astrocytoma
C. Medulloblastoma
D. Meningioma

36. A child presents with seborrheic dermatitis, lytic skull lesions, ear discharge and hepatosplenomegaly. What is the most likely diagnosis?
A. Leukemia
B. Lymphoma
C. Histiocytosis
D. Multiple myeloma

37. A 2 year old child presented with scattered lesions in skull. Biopsy revealed Langerhans giant cells. Most common marker associated with this condition is?
A. CD1a
B. CD3
C. CD208
D. CD68

38. L-asparaginase is useful in management of?
A. AML
B. ALL
C. CML
D. CLL

39. Most common site of extramedullary relapse of ALL is?
A. CNS
B. Lung
C. Liver
D. Testis

40. Noonan syndrome and NF-1 are associated with which hematological malignancy?
A. ALL
B. AML
C. JMML
D. CML

41. Most common facial tumour in children is?
A. Rhabdomyosarcoma
B. Basal cell carcinoma
C. Malignant melanoma
D. Squamous cell carcinoma

42. A 5 year old child with progressive ataxia, loss of weight, raised intracranial pressure and enhancing midline mass should be suspected to have which malignancy?
A. Neuroblastoma with cranial metastasis
B. Medulloblastoma
C. ALL with CNS involvement
D. Glioblastoma multiforme

43. A malignant tumour of childhood, that metastasizes to bones most often, is:
A. Wilms tumour
B. Neuroblastoma
C. Adrenal gland tumour
D. Granulose cell tumour of ovary

44. Chang staging is used for?
A. Neuroblastoma
B. Medulloblastoma
C. Wilms tumour
D. AML

45. Chloroma is commonly produced in children by?
A. AML
B. ALL
C. Neuroblastoma
D. Rhabdomyosarcoma

26. b	27. a	28. c	29. a	30. c	31. a	32. c	33. b	34. c	35. c
36. c	37. a	38. b	39. a	40. c	41. a	42. b	43. b	44. b	45. a

26. B. Fragile X syndrome

Fragile X syndrome is a trinucleotide repeat disorder but is not associated with malignancy. As per Nelson 20th Ed, all other options are associated with malignancy.

- Down syndrome: Increased risk of AML-M7, ALL
- Fanconi anemia: Increased risk of Leukemia, myelodysplastic syndrome, liver neoplasias etc.
- Bloom syndrome: Leukemia, lymphomas and various solid tumours

27. A. Hemangioma

Hemangiomas are the most common benign tumours in childhood. Leukemias are most common malignant tumour in children.

28. C. ALL

Malignancies in children

- MC: Leukemias (ALL)
- 2nd MC: Brain tumours (Most common solid malignancy)
- 3rd MC: Neuroblastoma (Most common extracranial solid malignancy)

29. A. Neuroblastoma

Neuroblastoma is a common malignancy in the first 2 years of life, presenting with abdominal mass and associated signs and symptoms. Metastatic disease can cause a variety of signs and symptoms, including fever, irritability, failure to thrive, bone pain, cytopenias, bluish subcutaneous nodules, orbital proptosis, and periorbital ecchymoses, producing the so called "raccoon eyes", as shown in the picture.

30. C. Retinoblastoma

Retinoblastomas are the most common inherited malignancy in children. Retinoblastoma can be either hereditary or sporadic. Hereditary cases usually are diagnosed at a younger age and are multifocal and bilateral, whereas sporadic cases are usually diagnosed in older children who tend to have unilateral, unifocal involvement. The hereditary form is associated with loss of function of the retinoblastoma gene (RB1) via gene mutation or deletion. The RB1 gene is located on chromosome 13q14 and encodes the retinoblastoma protein, a tumor suppressor protein that controls cell-cycle phase transition and has roles in apoptosis and cell differentiation.

31. A. Craniopharyngioma

Craniopharyngioma is a supratentorial tumour in children which arises as a suprasellar mass and produces suprasellar calcification.

32. C. Astrocytoma

Cerebellar astrocytomas are the most common posterior fossa tumours and are mostly benign in nature.

33. B. Wilms tumour

The child has aniridia, hemihypertrophy and genitourinary abnormalities. The most common associated malignancy in this child will be Wilms tumour.

- Syndromes associated with Wilms tumour: V important

Syndrome	Clinical Characteristics
Wilms tumor, aniridia, genitourinary abnormalities, and mental retardation (WAGR)	Aniridia, genitourinary abnormalities, mental retardation
Denys-Drash	Early-onset renal failure with renal mesangial sclerosis, male pseudohermaphroditism
Beckwith-Wiedemann	Organomegaly (liver, kidney, adrenal, pancreas) imacroglossia, omphalocele, hemihypertrophy

34. C. Histology

Histology is the most important prognostic factor in children with Wilms tumour. Histology plays a major role in risk stratification of WT. Absence of anaplasia is considered a favourable histologic finding. Presence of anaplasia is further classified as focal or diffuse, both of which are unfavourable histologic findings.

35. C. Medulloblastoma

Medulloblastoma is the most common malignant brain tumour in children. Astrocytoma is most common benign brain tumour in children, not malignant.

36. C. Histiocytosis

The findings are consistent with Langerhans cell histiocytosis (LCH). LCH has an extremely variable presentation. The skeleton is involved in 80% of patients and may be the only affected site, especially in children older than 5 yr of age. Bone lesions may be single or multiple and are seen most commonly in the skull in the form of lytic lesions. Approximately 50% of patients experience skin involvement at some time during the course of disease, usually as a hard-to-treat scaly, papular, seborrheic dermatitis of the scalp, diaper, axillary, or posterior auricular regions. The lesions may spread to involve the back, palms, and soles. The exanthem may be petechial or hemorrhagic, even in the absence of thrombocytopenia. Localized or disseminated lymphadenopathy is present in approximately 33% of patients. Hepatosplenomegaly

occurs in approximately 20% of patients. Bilateral exophthalmos may be seen. Otitis media is present in 30-40% of patients; deafness may follow destructive lesions of the middle ear.

37. A. CD1a

Findings are suggestive of Langerhans cell histiocytosis (LCH).

	Disease	Cellular Characteristics Of Lesions
LCH	Langerharis cell histiocytosis	Langerhans-like cells (CDla-positive, CD207-positive) with Birbeck granules (LCH cells)

38. B. ALL

According to Nelson 20th Ed, in the treatment of childhood ALL, initial therapy, termed remission induction, is designed to eradicate the leukemic cells from the bone marrow. During this phase, therapy is given for 4 wk and consists of vincristine weekly, a corticosteroid such as dexamethasone or prednisone, and usually a single dose of a long-acting, pegylated L-asparaginase preparation.

39. A. CNS

Most common sites of relapse in ALL

- MC: Bone marrow
- 2nd MC: CNS
- 3rd MC: Testis

40. C. JMML

Juvenile myelomonocytic leukemia (JMML) was previously known as Juvenile CML. According to Nelson 20th Ed, most patients with JMML have been found to have mutations that lead to activation of the RAS oncogene pathway including NF1 and PTPN11. Patients with neurofibromatosis type 1 and Noonan syndrome have a predilection for this type of leukemia, since they have germline mutations involved in RAS signaling.

41. A. Rhabdomyosarcoma

The most common Paediatric soft tissue sarcoma, rhabdomyosarcoma, accounts for approximately 3.5% of childhood cancers. These tumors may occur at virtually any anatomic site but are usually found in the head and neck (25%), orbit (9%), genitourinary tract (24%), and extremities (19%); retroperitoneal and other sites account for the remainder of primary sites. It is the most common facial tumour in the Paediatric age group.

42. B. Medulloblastoma

According to Nelson 20th Ed, the classic triad of headache, nausea, and vomiting as well as papilledema is associated with midline or infratentorial tumors. Since child has weight loss and progressive ataxia, it suggests a locally invasive infratentorial malignant tumour, commonly being medulloblastoma in children. Enhancement is further supportive of medulloblastoma. Remember that glioblastoma occurs in adults, not children.

43. B. Neuroblastoma

Neuroblastoma is the most common tumour metastasising to bones in children.

Also remember: As per Nelson 20th Ed, the most common sites of metastasis are the regional or distant lymph nodes, long bones and skull, bone marrow, liver, and skin. Lung and brain metastases are rare, occurring in >3% of cases.

44. B. Medulloblastoma

Chang staging is done for medulloblastomas

Chang Staging System for Medulloblastoma

T1	Tumor <3 cm in diameter
T2	Turn or >3 cm in diameter
T3a	Tumor >3 cm in diameter with extension into aqueduct of Sylvius or foramen of Luschka
T3b	Tumor >3 cm in diameter with unequivocal extension into the brainstem
T4	Tumor >3 cm in diameter with extension up past the aqueduct of Syfviusand/or down past the foramen magnum (ie; Beyond the posterior fossa}
MO	No evidence of subarachnoid or hematogenous metastasis
Ml	Turn or cells found in cereb nospi nal fluid
M2	Intracranial tumor beyond primary site
M3	Gross nodular seeding in spina) subarachnoid space
M4	Metastasis outside the cerebrospinal axis (especially to bone marrow, bone)

45. A. AML

Chloroma, also called granulocytic sarcoma, is an extramedullary manifestation of acute myeloid leukemia. It is a solid collection of leukemic cells occurring outside of the bone marrow.

46. According to latest NRP guidelines, in a vigorous term baby, cord clamping should be done?

A. Immediately at birth
B. After delay of 15 seconds
C. After delay of 30 to 60 seconds
D. After delay of 90 seconds

47. All the following are component of APGAR score except?

A. Muscle tone
B. Colour of the body
C. Heart rate
D. Respiratory rate

48. A baby delivered by emergency LSCS had meconium stained amniotic fluid. The baby did not cry immediately after birth, was hypotonic and heart rate was <100/min. Which will be the immediate treatment modality in this baby?

A. Starting positive pressure ventilation
B. Drying and stimulation
C. Suctioning the residual meconium from the hypopharnyx
D. Intubating the trachea and suctioning meconium from lower airway

49. A baby is born with meconium stained liquor. Which of the following is taken account of in terming a baby 'Vigorous' except?

A. Tone
B. Colour
C. HR
D. Respiration

50. A 1 minute old neonate with apnea is on bag and mask ventilation for last 30 seconds. He now has heart rate of 110 per minute and spontaneous breathing efforts. What next needs to be done?

A. Continue ventilation for 30 sec more
B. Continue ventilation for 60 sec more
C. Intubate and start ventilation
D. Discontinue ventilation

51. Absolute contraindication for bag and mask ventilation is?

A. Congenital diaphragmatic hernia
B. Meconium aspiration syndrome
C. Trachea-esophageal fistula
D. All of the above

52. Which of the following noted in a new born is non-innocuous?

A. Vaginal bleeding on day 4 of life
B. Central cyanosis
C. Erythematous papules in trunk and face on day 2
D. Bilateral subconjunctival haemorrhage

53. A neonate delivered at term was noticed to have multiple violet-black macules on the back and thigh since birth, as shown in the picture. Which among the following is not true regarding this condition?

A. A benign, self-limited transient phenomenon
B. May represent neural crest migration defect
C. May be associated with Hurler syndrome
D. Malignant degeneration in 1% cases

54. Retinopathy of prematurity is commonly predisposed by:

A. Less gestational age
B. Low birth weight
C. Oxygen toxicity
D. Carbohydrate excess

55. Least likely finding in neonatal sepsis is:

A. Increased C Reactive Protein
B. Neutrophilia
C. Increased ESR
D. Toxic granulated multilobulated nuclei

56. Most common cause of neonatal sepsis?

A. Streptococcus pneumoniae
B. Klebsiella
C. E coli
D. Pseudomonas

57. Most common cause of neonatal sepsis in India?

A. Klebsiella
B. Group B streptococci
C. E coli
D. Pseudomonas

58. In a preterm neonate admitted in ICU, which fungal pathogen can be transmitted from the hands of caregivers?

A. Candida albicans
B. Aspergillus fumigatus
C. Candida tropicalis
D. Candida parapsilosis

46. c	47. d	48. a	49. b	50. d	51. a	52. b	53. d	54. a	55. b
56. c	57. a	58. d							

59. IUGR babies are more prone to develop the following complications except:

A. Hypoglycemia
B. RDS
C. Polycythemia
D. Hypocalcemia

60. All the following can occur in a neonate for heat production except?

A. Shivering
B. Breakdown of brown fat with adrenaline secretion
C. Universal flexion like a foetus
D. Cutaneous vasoconstriction

61. Which mechanism of phototherapy is chiefly responsible for reduction in serum bilirubin?

A. Photooxidation
B. Photoisomerization
C. Structural isomerization
D. Conjugation

62. Most common pathological cause of neonatal jaundice?

A. Biliary atresia
B. Congenital hypothyroidism
C. Rh incompatibility
D. ABO incompatibility

63. A neonate born to O positive mother is found to be icteric on first day of life. What is the next step in management?

A. Start phototherapy
B. Start exchange transfusion
C. Send blood for total serum bilirubin measurement
D. Send blood to determine ABO and Rh-grouping

64. What is the normal disappearance time for the reflex being performed below?

A. 3 months
B. 5-6 months
C. 9-12 months
D. Persists throughout life

65. Abnormal persistence is defined if moro reflex is still present at?

A. Birth
B. 3 months
C. 5 months
D. 6 months

66. Neonatal reflex not present at birth?

A. Asymmetric tonic neck reflex
B. Symmetric tonic neck reflex
C. Moro reflex
D. Stepping reflex

67. A 1.5 kg child born at 32 weeks LSCS presents with moderate respiratory difficulty (RR 70/minutes). Which of the following is the immediate next management?

A. CPAP
B. Mechanical ventilation
C. Warm oxygen
D. Surfactant and ventilation

68. Drug of choice for prophylaxis of HMD in maternal therapy is?

A. Betamethasone
B. Dexamethasone
C. Prednisolone
D. Any potent corticosteroid can be used

69. Blood specimen for neonatal thyroid screening is obtained on-

A. Cord blood
B. 24 hr after birth
C. 48 hrs after birth
D. 72 hrs after birth

70. Which is not true regarding hypoxic ischemic encephalopathy?

A. In preterm babies with HIE, periventricular leukomalacia is the commonest lesion
B. Blood sugar should ideally be maintained in the hyperglycemic range in HIE babies
C. Corticosteroids and mannitol have no role in treatment of HIE
D. Phenobarbitone used at initial stage is associated with better long-term outcome.

71. A young neonate born at home and having history of delayed cry after birth is brought for poor feeding and seizures. His MRI shows the following abnormality (Red arrows).

Which among the following is the likely diagnosis in this child?

A. Periventricular leukomalacia
B. Germinal matrix haemorrhage
C. Status maromaratus with basal ganglia infarcts
D. Periventricular Inflammation with ventriculitis

59. b	60. a	61. c	62. c	63. a	64. b	65. d	66. b	67. c	68. b
69. d	70. b	71. a							

72. **Blood sugar estimation is relevant in all the following except?**
   A. Rh incompatibility
   B. Birth asphyxia
   C. SFD babies
   D. Baby of hypothyroid mother

73. **Most common cause of neonatal seizures?**
   A. HIE
   B. Intracranial HGE
   C. CNS infection
   D. Metabolic disease.

Q74. **Drug of choice for neonatal seizures is**
   A. Phenytoin
   B. Phenobarbitone
   C. Diazepam
   D. Sodium valproate

75. **All are features of neonatal necrotizing enterocolitis except-**
   A. Abdominal distension
   B. Increased bowel sounds
   C. Metabolic acidosis
   D. Pneumoperitoneum

76. **Radiological hallmark of NEC is?**
   A. Pneumatosis intestinalis
   B. Portal vein gas
   C. Ascites
   D. Perforation of the gut

77. **A nonventilated preterm baby in incubator is under observation. Which is the best way to monitor the baby's breathing and detect apnea?**
   A. Infrared thoracic movement study
   B. Capnography
   C. Nasal digital temperature monitoring
   D. Impedance technique

78. **Difference between seizure and jitteriness in a neonate all except-**
   A. Sensitivity to stimulus
   B. Frequency of movement
   C. Abnormality of gaze
   D. Autonomic disturbances.

79. **All of the following are complications in the new born of a diabetic mother except?**
   A. Hyperbilirubinemia
   B. Hyperglycemia
   C. Hypocalcemia
   D. Hypomagnesemia

80. **Infant of diabetic mother with weight 3.8 kg presented with seizures after 16 hours of birth. What is the cause?**
   A. Hypoglycemia
   B. Hypocalemia
   C. Birth asphyxia
   D. Intraventricular haemorrhage

81. **A child with respiratory distress and excess frothing from mouth is brought to emergency on day 2 of life. What is the most likely syndrome associated with this condition?**
   A. CHARGE syndrome
   B. Denys-drash syndrome
   C. VACTERL syndrome
   D. Russell-silver syndrome

82. **A neonate having congenital diaphragmatic hernia is brought with respiratory distress to emergency on 2nd day of life. On intubating, the apex beat shifted to right and distress worsened. What is the next step in management?**
   A. Insert nasogastric tube
   B. CXR to confirm tube position
   C. Perform capnography
   D. Emergency surgery

83. **Characteristic radiological feature of transient tachypnea of newborn is?**
   A. Reticulogranular appearance
   B. Low lung volume
   C. Air bronchogram
   D. Prominent horizontal fissure

84. **The most common sign of neonatal hypoxia is?**
   A. Tachycardia
   B. Bradycardia
   C. Ventricular arrhythmias
   D. Asystole

85. **All of the following are features of prematurity in a neonate except?**
   A. No creases on sole
   B. Abundant lanugo
   C. Thick ear cartilage
   D. Empty scortum

72. d	73. a	74. b	75. b	76. a	77. d	78. a	79. b	80. a	81. c
82. c	83. d	84. b	85. c						

46. C. After delay of 30 to 60 seconds

Current evidence suggests that cord clamping should be delayed for at least 30 to 60 seconds for most vigorous term and preterm newborns. If the placental circulation is not intact, such as after a placental abruption, bleeding placenta previa, bleeding vasa previa, or cord avulsion, the cord should be clamped immediately after birth. There is insufficient evidence to recommend an approach to cord clamping for newborns who require resuscitation at birth.

47. D. Respiratory rate

Apgar score

Sign	Score		
	1	2	3
Color	Pale blue	Pinlc body, blue extremities	Completely pink
Reflex irritability	None	Grimace	Vigorous cry
Heart rate	Absent	Slow {<100)	>100
Respiratory effort	Absent	Slow (irregular)	Crying
Muscle tone	Flaccid	Some flexion of extremities	Active motion

Score interpretation	
Score	Status
7-10	Normal
4-6	Moderately depressed
0-3	Severely depressed

48. A. Starting positive pressure ventilation

As per latest NRP guidelines, there is no role of routine tracheal suction and stimulation in non vigorous baby born through meconium stained liquor.

49. B. Colour

Criteria to define non vigorous baby: Any one of the following

- HR < 100 per min
- Poor resp effort
- Poor muscle tone

50. D. Discontinue ventilation

As per NRP guidelines, heart rate above 100 per min and spontaneous breathing efforts are an indication to stop ventilation and give post resuscitation care.

51. A. Congenital diaphragmatic hernia

CDH is absolute CI whereas MAS and TEF are relative CI for bag and mask ventilation

52. B. Central cyanosis

Non-innocuous means "not innocent", i.e. pathological. Central cyanosis is always pathological. However, other findings may be transiently seen even in normal neonates.

53. D. Malignant degeneration in 1% cases

This is a neonate showing Mongolian spots. They are a self-limited neural crest migration defect and occur in normal neonates, slowly fading with age. Extensive spots may occur in Hurler syndrome. They do not require any therapy.

54. A. Less gestational age

According to Nelson 20th Ed, the risk factors associated with ROP are not fully known, but prematurity and the associated retinal immaturity at birth represent the major factors. Oxygenation, respiratory distress, apnea, bradycardia, heart disease, infection, hypercarbia, acidosis, anemia, and the need for transfusion are thought by some to be contributory factors.

55. B. Neutrophilia

Neonatal sepsis shows neutropenia, not neutrophilia.

56. C. E coli

- MC Cause of neonatal sepsis in World/Overall: Group B strep (Strep agalactiae)> E.coli> Klebsiella
- MC Cause of neonatal sepsis in India: Klebsiella> Staph aureus> Pseudomonas> E coli
- MC fungal cause of neonatal sepsis (World and India): Candida parapsilosis

57. A. Klebsiella

See the explanation of Q11

58. D

See the explanation of Q11

59. B. RDS

Problems seen in IUGR babies include

- Asphyxia
- Meconium aspiration
- Polycythemia
- Hypoglycaemia
- Hypocalcemia
- Poor growth
- Increased risk of infections

However, hyaline membrane disease (HMD), also called respiratory distress syndrome (RDS) is not seen in these babies since they have adequate surfactant production due to in utero high cortisol production.

60. A

Newborns can not shiver. They show non-shivering thermogenesis, mediated by lipolysis of brown fat for heat production

61. C. Structural isomerization

Phototherapy (PTx) remains the mainstay of treating hyperbilirubinemia in neonates. PTx is highly effective and carries an excellent safety track record of over 50 years. It acts by converting insoluble bilirubin (unconjugated) into soluble isomers that can be

excreted in urine and feces. The most important mechanism is by Structural isomerization, where bilirubin gets converted into Lumirubin.

62. **C. Rh incompatibility**

Mostly the jaundice seen in neonates is physiological jaundice. However, hemolytic causes are the most common cause of pathological jaundice. Among hemolytic causes, Rh incompatibility is the single MC cause.

63. **A. Start phototherapy**

As per AIIMS protocol, in a neonate having serious jaundice, starting phototherapy is the first step in management, followed by investigating the child by sending blood for TSB measurement.

- Criteria for serious jaundice:
  a. Presence of visible jaundice in first 24 h
  b. Yellow palms and soles anytime
  c. Signs of acute bilirubin encephalopathy or kernicterus: hypertonia, abnormal posturing such as arching, retrocollis, ophisthotonus or convulsion, fever, high pitched cry)
  d. TcB (Transcutaneous bilirubinometer) value more than 95th centile as per age specific nomogram.

64. **B. 5-6 months**

This is asymmetric tonic reflex. On turning head to one side, there is extension on same side and flexion of limbs on opposite side. Disappears by 5-6 months of age.

65. **D. 6 months**

Moro reflex is a primitive neonatal reflex which normally appears before birth and tends to disappear between 3 to 6 months. Abnormal persistence is defined as persistence at 6 months or longer and mostly indicates frontal lobe damage.

66. **B. Symmetric tonic neck reflex**

In a term baby, all neonatal reflexes are present at birth, except **SPL**

- S: Symmetric tonic neck reflex (Cat's reflex)
- P: Parachute reflex
- L: Landau reflex

67. **C. Warm oxygen**

Warm humidified oxygen should be provided at a concentration initially sufficient to keep arterial oxygen pressure between 50 and 70 mm Hg (91-95% saturation) in order to maintain normal tissue oxygenation while minimizing the risk of oxygen toxicity. If oxygen saturation cannot be kept >90% at inspired oxygen concentrations of 40-70% or greater, applying CPAP at a pressure of 5-10 cm H2O via nasal prongs is indicated and usually produces a rapid improvement in oxygenation. CPAP reduces collapse of surfactant-deficient alveoli and improves both FRC and ventilation–perfusion matching.

68. **B. Dexamethasone**

According to latest guidelines, maternal administration of dexamethasone is the best strategy to attain early lung maturity to prevent HMD. The regimen comprises 4 doses od dexamethasone, 6 mg i.m. every 12 hours.

69. **D. 72 hrs after birth**

Screening for congenital hypothyroidism is done by heel pad prick, beyond 48 hr of life, ideally AT 72 HOURS. (Some texts give range as 2-4 days). Most common approach is TSH alone, followed by T4 if needed. Ideal approach is both TSH and T4 but is costly and not practically done in most centers.

70. **B. Blood sugar should ideally be maintained in the hyperglycemic range in HIE babies**

Both hypoglycaemia and hyperglycemia should be avoided in children with perinatal asphyxia. Target blood glucose in HIE is 70 to 100 mg/dl. Other options are correct.

71. **A. Periventricular leukomalacia**

This MRI is showing increased periventricular T2 signal on FLAIR image indicating PVL.

72. **D. Baby of hypothyroid mother**

Screening for neonatal hypoglycaemia is recommended in:

- Preterm, LBW, Small for date babies
- Birth asphyxia
- Rh incompatibility
- Infant of diabetic mother
- Polycythemia
- Nesidioblastosis
- Maternal drugs e.g. Labetalol

73. **A. HIE**

Anoxic damage leading to hypoxic ischemic encephalopathy (HIE) is the overall MC cause of neonatal seizures.

74. **B. Phenobarbitone**

Phenobarbitone is the drug of choice in neonatal seizures. The dose is 20 mg/kg/IV slowly over 20 minutes (not faster than 1 mg/kg/min).

75. **B. Increased bowel sounds**

Peristalsis is adversely affected in NEC and so these babies tend to develop decreased bowel sounds. A others can be seen.

76. **A. Pneumatosis intestinalis**

Pneumatosis intestinalis (air bubbles in the wall small intestine with gut wall edema) is the radiologic hallmark of necrotizing enterocolitis (NEC). This fir appears in Stage IIA of the modified Bell's staging f NEC.

77. **D. Impedance technique**

Best way to monitor respiration in an intubated ba is by capnography, whereas in a non-ventilated/nc intubated baby is by impedance technique, based up measuring the electrical resistance during breathi picked up by electrodes placed on the chest wall.

78. **A. Sensitivity to stimulus**

Seizures should be distinguished from the jitterin that may be present in normal newborns, in infa of diabetic mothers, in those who experienced b asphyxia or drug withdrawal, and in polycythe

neonates. An examiner may stop the jitteriness resembling simple tremors by holding the infant's extremity; this provides tactile stimulus and tends to briefly stop the jitteriness. Jitteriness often depends on sensory stimuli and occurs when the infant is active, and it is not associated with abnormal eye movements.

79. B. Hyperglycemia

Hypoglycemia occurs in infants of diabetic mothers due to fetal hyperinsulinemia. **Hypoglycemia** develops in approximately 25-50% of infants of diabetic mothers and 15-25% of infants of mothers with gestational diabetes, but only a small percentage of these infants become symptomatic.

80. A. Hypoglycemia

Cause of seizure in infant of diabetic mother

- First 24 to 36 hr: Hypoglycemia
- Beyond this period: Hypocalcemia

81. C. VACTERL syndrome

The symptoms are suggestive of tracheo-esophageal fistula with esophageal atresia. Fifty percent of infants are nonsyndromic without other anomalies, and the rest have associated anomalies, most often associated with the VATER or VACTERL (vertebral, anorectal, [cardiac], tracheal, esophageal, renal, radial, [limb]) syndrome. Cardiac and vertebral anomalies are seen in 32% and 24%, respectively. These syndromes generally are associated with normal intelligence.

82. C. Perform capnography

In a child with CDH, bag and mask ventilation is contraindicated. If a child with CDH is asymptomatic, first clinical intervention is to insert a nasogastric tube and keep it open. If however he presents in distress (as in this question), the first clinical intervention is to intubate the child and start PPV by ET tube. Now if distress worsens and apex beat shifts to right, it suggests esophageal intubation, which can be confirmed by capnography for ETCO2 measurement. If capnography not available, remove tube and reattempt intubation. There is no role of emergency surgery in CDH.

83. D. Prominent horizontal fissure

According to Nelson 20th Ed, the chest radiograph in TTN shows prominent pulmonary vascular markings, fluid in the intralobar fissures, overaeration, flat diaphragms, and, rarely, small pleural effusions. Features suggestive of HMD like reticulogranular pattern, air bronchogram and low lung volume are not seen.

84. B. Bradycardia

Most common response of fetus and neonates to hypoxia is bradycardia.

85. C. Thick ear cartilage

Features of preterm neonates:

- Birth weight and length is less
- Undescended testis in males and widely separated labia in females
- Poorly developed or absent creases on sole of the foot
- Abundant vernix caseosa
- Abnormal irregular breathing
- Lanugo hair
- Thin, soft, compliant and poorly developed ear cartilage
- Thin, transparent skin with visible veins
- Absence of subcutaneous fat
- Poor neonatal reflexes

86. A 6-week-old male infant presents with projectile vomiting since past 5 days. Physical examination reveals a dehydrated, irritable infant. Laboratory evaluation reveals hypochloremic, hypokalemic metabolic acidosis. The likely diagnosis is?

A. Physiological reflux
B. Hypertrophic pyloric stenosis
C. Duodenal atresia
D. Hirschsprung's disease

87. Most common cause of diarrhea in infant is?

A. Bacteria
B. Virus
C. Formula feeds
D. Protozoa

88. Most common bacterial cause of diarrhea in children is?

A. E.coli
B. Salmonella
C. Staphylococcus
D. Shigella

89. One of the intestinal enzymes that is generally deficient in children following an attack of severe infectious enteritis is?

A. Lactase
B. Trypsin
C. Lipase
D. Amylase

90. Most common cause of GI Tract obstruction in children aged 1 ½ years is?

A. Volvulus
B. Intussusception
C. Pyloric stenosis
D. Intestinal atresia

91. Most common cause of GI obstruction in neonate is?

A. Meconium ileus
B. Meconium plug syndrome
C. Duodenal atresia
D. Jejunal atresia

92. A 9 month old child is passing 10 to 12 loose stools per day for last 9 days. There is no blood or mucus in stools. There is no dehydration as well. Which is the therapy of choice in such a child?

A. ORS alone
B. ORS with Zinc
C. ORS with zinc with antibiotics
D. ORS with zinc with probiotics

93. The histopathology of jejunal biopsy in a child with celiac disease shows all except?

A. Crypt hyperplasia
B. Thickened mucosa
C. Decreased villi: crypt ratio
D. Increased lymphocytes in the lamina propria

94. Molar ratio of sodium and glucose in WHO low osmolarity ORS is?

A. 1:1
B. 1:2
C. 2:3
D. 2:5

95. Ramu, a 8-year old boy presents with upper GI bleeding. On examination he is found to have splenomegaly: there are no sign of ascites, or hepatomegaly. Esophageal varices are found on UGIE. Most likely diagnosis is?

A. Budd-Chiari syndrome
B. Non-cirrhotic portal fibrosis
C. Cirrhosis
D. Veno-occlusive disease

96. A neonate has not passed meconium beyond 48 hours of life. What is the next investigation to perform?

A. Chest X ray
B. Babygram
C. Ultrasound abdomen
D. Contrast enema

97. Dose of zinc approved for use in childhood diarrhea by WHO is?

A. 5 mg/day
B. 10 mg/day
C. 20 mg/day
D. 50 mg/day

98. Most common cause of neonatal cholestasis is?

A. Idiopathic neonatal hepatitis
B. Extrahepatic biliary atresia
C. Choledochal cyst
D. Wilson disease

99. Test useful to distinguish neonatal hepatitis and biliary atresia is?

A. GGT
B. 5'- Nucleotidase
C. Alkaline phosphatase
D. Ultrasound

100. Most reliable test to diagnose biliary atresia is?

A. USG abdomen
B. Percutaneous liver biopsy
C. HIDA scan scintigraphy
D. Peroperative cholangiogram

101. The radiological sign seen below is suggestive of a disease, whose clinical hallmark is?

A. Abdominal distension
B. Bilious vomiting
C. Constipation
D. Jaundice

86. b	87. b	88. a	89. a	90. b	91. b	92. b	93. b	94. a	95. b
96. d	97. c	98. a	99. d	100. b	101. b				

102. A mother asks if a 3-month child can be started on cereals. If done, this child will be at risk for which among the following?

A. Retarded oral development
B. Food allergy
C. Risk of increased bacterial infections of the gut
D. Increased reflux

103. A 3-year-old child presented with acute onset of abdominal pain, vomiting and progressive lethargy. Contrast enema is shown in the picture. What is the likely diagnosis?

A. Volvulus
B. Intussusception
C. Meckel's diverticulitis
D. Adhesions

104. Which of the following is the earliest indicator of pathological gastroesophageal reflux in infants (GERD)

A. Respiratory symptoms
B. Postprandial regurgitation
C. Upper GI Bleed
D. Stricture esophagus

105. Most common indication for liver transplantation in children is?

A. Viral hepatitis with fulminant hepatic failure
B. Biliary atresia
C. Metabolic disease
D. Hepatic tumours

102. b	103. b	104. a	105. b

86. B. Hypertrophic pyloric stenosis

This is the classic presentation of congenital or idiopathic hypertrophic pyloric stenosis. Mostly presents between 6 week to 3 month age in first born males with projectile vomiting post-feed. Dehydration with hypochloremic alkalosis and mild to moderate hypokalemic is commonly seen.

87. B. Virus

Most common cause of diarrhea in infants and young children is viral infection, most commonly Rotavirus.

88. A. E.coli

Most common bacterial cause of diarrhea in children is E.coli (Enteropathogenic E.coli, EPEC).

89. A. Lactase

Young children with acute diarrhea, typically due to infectious gastroenteritis, may temporarily stop producing lactase, the intestinal enzyme that digests lactose. Thus, they may not digest lactose, the main sugar in milk, and this may worsen or prolong the diarrheal illness, eventually leading to persistent diarrhea.

90. B. Intussusception

Intussusception occurs when a portion of the alimentary tract is telescoped into an adjacent segment. It is the most common cause of intestinal obstruction between 5 mo and 3 yr of age and the most common abdominal emergency in children younger than 2 yr.

91. B. Meconium plug syndrome

Most common causes of neonatal obstruction

- MC: Meconium plug syndrome
- 2nd MC: Duodenal atresia
- 3rd MC: Hirschsprung disease
- 4th MC: Meconium ileus due to cystic fibrosis

92. B. ORS with Zinc

This child has acute watery diarrhea and no dysentery. Hence needs only ORS with Zinc. Antibiotics are indicated only in dysentery. Probiotics are not routinely recommended in childhood diarrhea as per WHO.

93. B. Thickened mucosa

Mucosa is thinned out with subtotal to total villous atrophy in celiac disease. Other findings are seen in these patients.

Findings seen on biopsy in celiac disease include

- Increased intraepithelial lymphocytes in gastric mucosal epithelium and submucosa
- Crypt hyperplasia with a decreased villi: crypt ratio
- Blunted or atrophic villi
- Mononuclear cell infiltration in the lamina propria

94. A. 1:1

Composition of Standard and Reduced Osmolarity ORS

	Standard ORS	Reduced (mEq or mmol/l) Osmolarity ORS
Glucose	111	75
Sodium	90	75
Chloride	80	65
Potassium	20	20
Citrate	10	10
Osmolarity	311	245

95. B. Non-cirrhotic portal fibrosis

This child has esophageal varices with splenomegaly but no hepatomegaly or jaundice. This favours a diagnosis of NCPF. Budd chairi syndrome will have hepatomegaly and jaundice and so will veno-occlusive disease have. Cirrhosis has varices with ascites and jaundice.

96. D. Contrast enema

In delayed passage of meconium, neonatal intestinal obstruction is suspected and the investigation of choice is Water soluble contrast enema. It is both diagnostic and therapeutic.

97. C. 20 mg/day

All children older than 6 months of age with acute diarrhea in at-risk areas should receive oral zinc (20 mg/ day) in some form for 10-14 days.

98. A. Idiopathic neonatal hepatitis

Neonatal cholestasis is defined biochemically as prolonged elevation of the serum levels of conjugated bilirubin beyond the 1st 14 days of life. Most common cause in neonates is idiopathic neonatal hepatitis, followed by extrahepatic biliary atresia.

99. D. Ultrasound

Abdominal ultrasound is a helpful diagnostic tool in evaluating neonatal cholestasis. The gallbladder either is not visualized or is a micro-gallbladder in patients with biliary atresia. Children with intrahepatic cholestasis caused by idiopathic neonatal hepatitis, cystic fibrosis, or total parenteral nutrition can have similar ultrasonographic findings. Ultrasonographic triangular cord sign, which represents a cone-shaped fibrotic mass cranial to the bifurcation of the portal vein, may be seen in patients with biliary atresia.

100. B. Percutaneous liver biopsy

According to Nelson 20th Ed, Percutaneous liver biopsy is the most valuable procedure in the evaluation of neonatal hepatobiliary diseases and provides the most reliable discriminatory evidence. Biliary atresia is characterized by bile ductular proliferation, the presence of bile plugs, and portal or peri-lobular edema and fibrosis, with the basic hepatic lobular architecture intact. Hepatobiliary scintigraphy with technetium-labelled iminodiacetic acid derivatives

(e.g. HIDA) is a sensitive but not specific test for biliary atresia. Peroperative cholangiogram is a highly specific test but is invasive and done during surgical laparotomy.

101. B. Bilious vomiting

This is the abdominal radiograph of a newborn infant held upright. Note the "double-bubble" gas shadow above and the absence of gas in the distal bowel in this case of congenital duodenal atresia. According to Nelson 20th Ed, the hallmark of duodenal obstruction is bilious vomiting without abdominal distention, which is usually noted on the 1st day of life. Peristaltic waves may be visualized early in the disease process. A history of polyhydramnios is present in half the pregnancies and is caused by inadequate absorption of amniotic fluid in the distal intestine. This fluid may be bile stained because of intrauterine vomiting. Jaundice is present in one-third of the infants.

102. B. Food allergy

According to Nelson 20th Ed, starting cereals before 4 months of age increases the risk of sensitization to food-borne allergens and predisposes to increased risk of food allergies.

103. B. Intussusception

In typical cases of intussusception, there is sudden onset, in a previously well child, of severe paroxysmal colicky pain that recurs at frequent intervals and is accompanied by straining efforts with legs and knees flexed and loud cries. At times, the lethargy is disproportionate to the abdominal signs. Eventually, a shock-like state, with fever and peritonitis, can develop. Vomiting occurs in most cases and is usually more frequent in the early phase. In the later phase, the vomitus becomes bile stained. 60% of infants pass a stool containing red blood and mucus, the currant jelly stool. The diagnostic findings of intussusception on ultrasound include a tubular mass in longitudinal views and a doughnut or target appearance in transverse images. Contrast enemas demonstrate a filling defect or cupping in the head of the contrast media where its advance is obstructed by the intussusceptum. A central linear column of contrast media may be visible in the compressed lumen of the intussusceptum, and a thin rim of contrast may be seen trapped around the invaginating intestine in the folds of mucosa within the intussuscipiens (coiled-spring sign), especially after evacuation.

104. A. Respiratory symptoms

Postprandial regurgitation can occur as normal reflux too. Pathological reflux is indicated by presence of respiratory symptoms with features of reflux.

105. B. Biliary atresia

As Nelson 20th Ed says "Biliary atresia is the most common indication for liver transplantation in children, followed by metabolic and inborn disorders, autoimmune and familial cholestatic disorders, and acute hepatic necrosis."

106. Most common congenital anomaly of larynx is?

A. Laryngeal web
B. Laryngeal stenosis
C. Laryngomalacia
D. Laryngeal papilloma

107. Chandu, a one-year old child with pneumonia should be admitted in hospital for the following complaints except:

A. Refusal of feed
B. Persistent vomiting
C. Fever 39° C
D. Difficulty in waking

108. A 6-year old boy is brought to the emergency room with 3-hour history of fever to 100°F & sore throat. The child appears alert but anxious and toxic. He has mild inspiratory stridor and is drooling. You should immediately-

A. Examine the throat and obtain a culture
B. Obtain ABG and start on IV line
C. Order a chest X-ray and lateral view of neck
D. Prepare to establish an airway

109. A 2-year old boy came to emergency department at 3 am with dyspnoea and fever. It was associated with barking cough and stridor was present only on crying. But child was consolable and well hydrated and was able to drink. Next management is –

A. Nebulized epinephrine (racemic)
B. Dexamethasone
C. Blood culture and blood examination
D. Nasal washings of RSV and influenza virus

110. A 3-year old boy is brought to the casualty by his mother with progressive shortness of breath for 1-day duration. The child has history of bronchial asthma. On examination the child is blue, gasping and unresponsive. What will you like to do first–

A. Intubate and ventilate
B. Administer 100% oxygen by mask
C. Give hydrocortisone injection immediately
D. Start salbutamol nebulisation

111. A 2-year old female child was brought to a PHC with a history of cough and fever for 4 days with inability to drink for last 12 hours. On examination, the child was having weight of 5 kg and RR of 45/min with fever. The child will be classified as suffering from which among the following as per latest WHO categorisation?

A. Very severe pneumonia
B. Severe pneumonia with danger sign
C. Pneumonia
D. No pneumonia

112. All of the following are seen in the viral pneumonia except

A. Presence of interstitial inflammation
B. Predominance of alveolar exudates
C. Association with bronchiolitis
D. Multinucleate giant cells in the bronchiolar wall

113. A newborn baby becomes cyanosed on closing its mouth which disappears on crying. The most probable diagnosis is

A. Tracheo esophageal fistula
B. GERD
C. Laryngomalacia
D. Bilateral choanal atresia

114. Raju, a one-year old child suffering from mild fever for 2 days associated with sneezing and running nose is brought with increasing respiratory distress. Two other family members had similar symptoms. On examination he has a respiratory rate of 45/minute, with nasal flaring, SCR, ICR and bilateral wheeze. The most likely diagnosis is –

A. Bronchiolitis
B. Viral croup
C. Asthma
D. Foreign body aspiration

115. A 2-year old child is labelled as having pneumonia according to WHO guidelines, if the child has respiratory rate greater than?

A. 30 per min
B. 40 per min
C. 50 per min
D. 60 per min

116. Most common cause of empyema in a child aged 2 years is?

A. E. coli
B. Pneumococcus
C. Klebsiella
D. Staphylococcus

117. A child aged 2 years with fever since 3 days and cough since one day has progressive respiratory distress and noisy breathing. X-ray neck appearance is shown below. What is the likely etiological agent?

A. Strep pneumoniae
B. Strep pyogenes
C. Parainfluenza virus
D. Hemophilus influenzae

106. c	107. c	108. d	109. a	110. a	111. b	112. b	113. d	114. a	115. b
116. d	117. c								

118. A child presented with recurrent sinusitis, situs inversus and dextrocardia. Likely diagnosis is?

A. Kartagener syndrome
B. William-Campbell syndrome
C. Ehlers-Danlos syndrome
D. Marfan syndrome

119. A 7 year old child presented with non-productive cough and dyspnea. After giving oral antibiotics, there was initial mild improvement, followed by increasing respiratory distress, mild fever and bilateral wheeze. Chest X-ray showed hyperlucency and PFT showed obstructive pattern. Most probable diagnosis is?

A. Follicular bronchitis
B. Bronchiolitis obliterans
C. Post-viral syndrome
D. Pulmonary alveolar microlithiasis

120. Most common cause of pneumonia in cystic fibrosis in a 3 year old child is?

A. Pseudomonas auruginosa
B. Mycoplasma pneumoniae
C. Burkholderia cepacian
D. Staphylococcus aureus

121. For a child with aspirated foreign body, treatment of choice is?

A. Chest X ray
B. Rigid bronchoscopy
C. Flexible bronchoscopy
D. Direct laryngoscopy

122. The most important cause of under 5 mortality rate is?

A. Diarrhea
B. Malnutrition
C. Respiratory infections
D. Trauma

123. Kumar 1½ year old child accidentally drinks kerosene mistaking it for water. After 6 hours he developed cough,tachypnea and retractions. In the emergency department, the first step in management is

A. Stimulate vomiting
B. Give charcoal
C. Do nasogastric lavage
D. Connect pulse oximeter & perform ABG if needed.

124. A previously healthy active 18-month old presents with unilateral nasal obstruction & foul-smelling discharge. The examination is otherwise unremarkable. The most likely diagnosis is

A. Foreign body
B. Nasal polyps
C. Frontal sinusitis
D. Choanal atresia

125. Staccato paroxysms of cough are seen in?

A. Allergic cough sinusitis
B. Tracheitis
C. Psychogenic cough
D. Chlamydia infection

126. An 18 month old child comes to a subcentre with fever and cough. On examination, weight was 11.5 kg, respiratory rate was 46/min and no chest indrawing. Most appropriate management?

A. IV antibiotics at PHC and observe
B. Give first dose antibiotic and refer to higher center
C. Oral antibiotics for 5 days and send home
D. Home based local remedies

127. Loeffler's syndrome is characterised by?

A. Transient, migratory pulmonary infiltrates
B. Fibrosis in lung apex
C. Military mottling
D. Non migratory transient infiltrates

128. Most common cause of ARDS in children is?

A. Aspiration
B. Injury
C. Severe pneumonia with sepsis
D. DIC

129. Pneumatoceles are produced by infection in children by which organism?

A. Staphylococcus aureus
B. Pseudomonas
C. Strep pyogenes
D. Hemophilus influenzae

130. The commonest cause of Bacterial pneumonia in children is:

A. Streptococcus pyogenes
B. Hemophilus influenza
C. Streptococcus pneumonia
D. Staphylococcus aureus

118. a	119. b	120. d	121. b	122. c	123. d	124. a	125. d	126. c	127. a
128. c	129. a	130. c							

106. C. Laryngomalacia

As per all Paediatric texts, laryngomalacia is the most common congenital anomaly of larynx.
Also remember: Laryngomalacia is the most common cause of intermittent stridor below 2 years of age.

107. C. Fever 39° C

Indications for hospital admission in childhood pneumonia (Ref. WHO update 2015, Nelson 20th Ed, PGI Paediatric protocols by Singhi S et al)

- Inability to feed or drink
- Lethargy: Abnormally sleepy or difficult to awake
- Persistent vomiting and dehydration
- Convulsions or seizures
- Age below 1 year
- Central cyanosis
- Severe grunting
- Severe malnutrition
- Suspected complication of pneumonia

108. D. Prepare to establish an airway

The child has features of acute epiglottitis. Immediately securing airway is paramount and should not be delayed pending investigations. After securing airway by intubation, we can start IV fluids, IV antibiotics and perform necessary investigations to confirm the diagnosis.
According to Nelson 20th Ed, "**Epiglottitis** is a medical emergency and warrants immediate treatment with an **artificial airway** placed under controlled conditions, either in an operating room or intensive care unit."

109. A. Nebulized epinephrine (racemic)

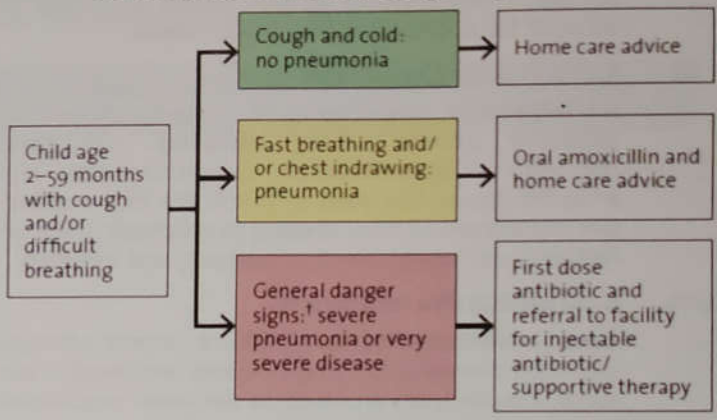
The child has acute laryngo-tracheo-bronchitis, i.e. Croup. Since child has stridor on crying, we label it as "moderate croup" which needs racemic epinephrine neobulization.
As per Nelson 20th Ed, "**Nebulized racemic epinephrine** is an accepted treatment for moderate or severe croup. The mechanism of action is believed to be constriction of the precapillary arterioles through the β-adrenergic receptors, causing fluid resorption from the interstitial space and a decrease in the laryngeal mucosal edema." Use of dexamethasone is reserved for mild croup.

110. A. Intubate and ventilate

Normally, any child presenting with acute severe asthma needs to be started on 100% oxygen followed by salbutamol nebulization. However, this child is blue, gasping and unresponsive (indicating almost complete airway obstruction with depressed consciousness). These children have high mortality and since respiratory efforts are poor (gasping), the child will not benefit from oxygen alone. We will immediately start bag and mask ventilation, followed by intubating the child to save the life of the child.
Also remember: In any child with severe persistent asthma without above features, starting 100% O2 will be the first step in management.

111. B. Severe pneumonia with danger sign

According to the updated WHO-2015 guidelines, the term very severe pneumonia has been replaced by severe pneumonia with danger signs.


Child age 2–59 months with cough and/or difficult breathing
Cough and cold: no pneumonia
Home care advice
Fast breathing and/or chest indrawing: pneumonia
Oral amoxicillin and home care advice
General danger signs:† severe pneumonia or very severe disease
First dose antibiotic and referral to facility for injectable antibiotic/ supportive therapy

* Not able to drink, persistent vomiting, convulsions, lethargic or unconscious, stridor in a calm child or severe malnutrition.

- Since this child has tachypnea (fast breathing) with danger signs like malnutrition (weight only 5 kg at 2 yr) and inability to drink for 12 hours, diagnosis is severe pneumonia with danger signs.

112. B. Predominance of alveolar exudates

Alveolar exudates are predominantly seen in bacterial pneumonias. Viral pneumonias have interstitial infiltrates, moderate grade fever and sometimes giant cells in the bronchiolar wall (esp in measles). RSV-associated pneumonia can have overlapping features with acute bronchiolitis.

113. D. Bilateral choanal atresia

The child is having cyclical cyanosis, suggestive of bilateral choanal atresia

114. A. Bronchiolitis

The child has features of respiratory distress with fever and bilateral wheeze. Any child below 2 yr with such illness is presumed to have acute bronchiolitis unless proven otherwise. Also remember:
As per Nelson 20th Ed, "**Acute bronchiolitis** is predominantly a viral disease. RSV is responsible for more than 50% of cases. Other agents include parainfluenza, adenovirus, rhinovirus, and *Mycoplasma*. Emerging pathogens include human metapneumovirus and human bocavirus, which may be a primary cause of viral respiratory infection or occur as a coinfection with RSV."

115. B. 40 per min

As per WHO and IMNCI

Age	Respiratory Rate (bpm)
Birth to 2 months	> 60
2-12 months	> 50
1-5 years	> 40

116. D. Staphylococcus

Staph aureus is the most common cause of empyema in young children upto 2 to 3 yr of age. In older children Strep pyogenes is more common.

117. C. Parainfluenza virus

The child is having "steeple sign" suggestive of subglottic narrowing, seen in croup. As per Nelson 20th Ed, "Parainfluenza viruses (types 1, 2, and 3) account for approximately 75% of cases."

118. A. Kartagener syndrome

Kartagener's syndrome is a rare, autosomal recessive genetic ciliary **disorder** comprising the triad of situs inversus, chronic sinusitis, and bronchiectasis. The basic problem lies in the defective movement of cilia, leading to recurrent chest infections, ear/nose/throat symptoms, and infertility.

119. B. Bronchiolitis obliterans

Bronchiolitis obliterans (BO), a chronic obstructive lung disease of the bronchioles and smaller airways, results from an insult to the lower respiratory tract leading to fibrosis of the small airways. In the nontransplant patient, BO most commonly occurs in the Paediatric population after respiratory infections, particularly adenovirus. Cough, fever, cyanosis, dyspnea, chest pain, and respiratory distress followed by initial improvement may be the initial signs of BO. Chest radiographs may be relatively normal compared with the extent of physical findings but can demonstrate hyperlucency and patchy infiltrates. PFT shows obstructive pattern. Exercise testing shows reduced exercise capacity andimpaired oxygen consumption. Ventilation–perfusion scans reveal a typical moth-eaten appearance of multiple matched defects in ventilation and perfusion. High-resolution chest CT often demonstrates patchy areas or a mosaic pattern of hyperlucency, air trapping, and bronchiectasis. No definitive therapy exists for BO. Administration of corticosteroids may be beneficial.

120. D. Staphylococcus aureus

Although MCC of pneumonia in cystic fibrosis is Pseudomonas aeruginosa in adults, however the most common cause below 12 yr is Staph aureus in cystic fibrosis. Ref. Nelson 20th Ed

121. B. Rigid bronchoscopy

As per Nelson 20th Ed, "the treatment of choice for airway foreign bodies is prompt endoscopic removal with rigid instruments", i.e. Rigid Bronchoscopy. It is both diagnostic and therapeutic.

122. C. Respiratory infections

As per WHO update, Pneumonia is the MCC of death below 5 yr of age, i.e. U5MR

123. D. Connect pulse oximeter & perform ABG if needed

Since kerosene is a volatile poison, aspiration pneumonitis can develop even with ingestion without obvious aspiration. These children need monitoring for early development of pneumonia. Gastric lavage and inducing vomiting should be avoided.

124. A. Foreign body

Unilateral nasal obstruction with foul-smelling discharge is the classic presentation of retained foreign body in the nose in young children.

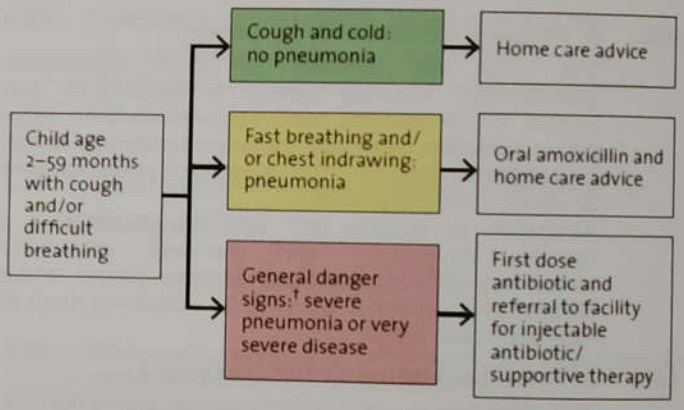
According to Nelson 20th Ed, "Unilateral nasal discharge and obstruction should suggest the presence of a foreign body, which can often be seen on examination with a nasal speculum or wide otoscope placed in the nose."

125. D. Chlamydia infection

As per Nelson 20th Ed and OP Ghai 8th Ed, Staccato paroxysms of cough are seen in chlamydia and whooping cough infections. Whereas honking cough is seen in psychogenic cough.

126. C. Oral antibiotics for 5 days and send home

Since child has tachypnea but no danger sign, diagnosis is pneumonia and oral antibiotics are sufficient.


Child age 2–59 months with cough and/or difficult breathing
Cough and cold: no pneumonia
Home care advice
Fast breathing and/or chest indrawing: pneumonia
Oral amoxicillin and home care advice
General danger signs:† severe pneumonia or very severe disease
First dose antibiotic and referral to facility for injectable antibiotic/ supportive therapy

127. A. Transient, migratory pulmonary infiltrates

Loeffler syndrome is characterized by transient, migrating lung infiltrates and by eosinophilia in the peripheral blood. It occurs in sensitized hosts during the stage of larval migration through the lungs. This syndrome can be a response to several parasitic infestations, **ascariasis** being the commonest aetiology.

128. C. Severe pneumonia with sepsis

Pneumonia with associated sepsis is the most common cause of ARDS in children as per all standard texts.

129. A. Staphylococcus aureus

Causes of pneumatocele: Staph aureus> Klebsiella. Ref. OP Ghai 8th Ed

130. C. Streptococcus pneumonia

Beyond 2 months of age, Strep pneumoniae is the most common bacterial cause of pneumonia in children.

Also remember: The causes of childhood pneumonia

Age Group	Frequent Pathogens (In Order Of Frequency)
Neonates (<3 wk)	Group B streptococcus, Escherichia coli, other Gram-negative bacilli, Streptococcus pneumoniae, Haemophilus influenzae (type b,* nontypeable)
3 wk-3 mo	Respiratory syncytial virus, other respiratory viruses (rhinoviruses, parainfluenza viruses, influenza viruses, adenovirus), S. pneumoniae, H. influenzae (type b,* nontypeable); if patient is afebrile, consider Chlamydia trachomatis
4 mo–4 yr	Respiratory syncytial virus, other respiratory viruses (rhinoviruses, parainfluenza viruses, influenza viruses, adenovirus), S. pneumoniae, H. influenzae (type b,* nontypeable), Mycoplasma pneumoniae, group A streptococcus
>5 yr	M. pneumoniae, S. pneumoniae, Chlamydophila pneumoniae, H. influenzae (type b,* nontypeable), influenza viruses, adenovirus, other respiratory viruses, Legionella pneumophila

Growth and Development, Nutrition — Questions

131. Fetal growth is maximally affected by?
A. Insulin
B. Cortisol
C. Growth hormone
D. Thyroxine

132. Tripling of weight occurs at what age?
A. 5 months
B. 6 months
C. 12 months
D. 24 months

133. Half the adult height is attained at what age?
A. 1 yr
B. 2 yr
C. 3 yr
D. 4 yr

134. The percentage gain in height at end of first year of life is?
A. 25%
B. 50%
C. 75%
D. 100%

135. The recommended dietary allowance (RDA) for zinc in age group of 1 to 3 years is?
A. 20 mg/day
B. 10 mg/day
C. 7.5 mg/day
D. 5 mg/day

136. Delayed dentition is failure of teeth to erupt by?
A. 6 months
B. 9 months
C. 12 months
D. 13 months

137. Long, mildly pigmented pubic hair with enlarged, pink scrotum is seen in which SMR stage in boys?
A. SMR 1
B. SMR 2
C. SMR 3
D. SMR 4

138. First sign of puberty in females?
A. Growth spurt
B. Menarche
C. Pubarche
D. Thelarche

139. First visible sign of puberty in females?
A. Growth spurt
B. Menarche
C. Pubarche
D. Thelarche

140. Microcephaly is defined as?
A. HC below 34 cm
B. HC below -2 SD
C. HC below -3 SD
D. HC below 10th centile

141. The important fatty acid present in breast milk, important for growth is –
A. Docosahexaenoic acid
B. Palmitic acid
C. Linoleic acid
D. Linolenic acid

142. Most common cause of short stature is?
A. Constitutional
B. Hypothyroidism
C. Growth hormone deficiency
D. Systemic illness

143. Which among the following is true about constitutional delay in growth?
A. Neonates with constitutional delay show birth anomalies
B. IGF-1 levels are low for chronological age
C. Bone age is normal
D. IGF-1 levels are low for bone age

144. Artificial feeding causes all of the following except?
A. NEC
B. Eczema
C. Late onset tetany
D. Hemorrhagic disease

145. A 6 year old child is brought to OPD with complaints that "the child doesn't eat much and is weak". On anthropometric assessment, weight of the child is found to be 18 kg and height of the child is 110 cm. What shall be your advice to the parents?
A. Start multivitamins with iron
B. Start calorie and fat rich diet
C. Start protein rich diet
D. Continue normal feeding as before

146. Best indicator for nutritional status for a child is?
A. Mid arm circumference
B. Head circumference
C. Rate of increase of height and weight
D. Chest circumference

147. Marker for both acute and chronic malnutrition is?
A. Weight for height
B. BMI
C. Weight for age
D. Height for age

148. All of the following conditions are observed in marasmus except-
A. Hepatomegaly
B. Muscle wasting
C. Low insulin levels
D. Extreme weakness

149. All of the following are features of kwashiorkor except –
A. High blood osmolarity
B. Hypoalbuminemia
C. Edema
D. Fatty liver

150. Diagnosis of kwashiorkor is based on?
A. Edema and skin changes
B. Edema and mental changes
C. Weight loss and mental changes
D. Bitot spots and hepatomegaly

131. a	132. c	133. b	134. b	135. d	136. d	137. b	138. a	139. d	140. c
141. a	142. a	143. b	144. d	145. d	146. c	147. c	148. a	149. a	150. b

151. Age independent anthropometric indices include all except:
A. Rao scale
B. Jellife index
C. Gomez index
D. Quac Stick

152. Cerebellar ataxia is feature of which vitamin deficiency?
A. Vit A
B. Vit C
C. Vit E
D. Vit D

153. When can a child speak future tense?
A. 2 years
B. 3 years
C. 4 years
D. 5 years

154. A child gets a vocabulary of 50 words at
A. 12 months
B. 15 months
C. 18 months
D. 24 months

155: Which among the following is a red flag sign for abnormally delayed development in a child?
A. Absence of vocalization by 6 months
B. Absence of pincer grasp by 10 months
C. Inability to stand without support by 12 months
D. Lack of imaginative play by 2 years

156. A child learns to draw a circle by what age?
A. 12 months
B. 24 months
C. 30 months
D. 36 months

157. A female child has recently learnt to eat with spoon without spilling, dress and undress under supervision and understand she is a girl. These skills are first acquired between the age of?
A. 2 and 3 years
B. 3 and 4 years
C. 4 and 5 years
D. 5 and 6 years

158. A child can self-feed with spoon without spilling, copy a circle, dress and undress independently but cannot close and open his buttons. What is the likely age at which this development status is first attained?
A. 2 years
B. 3 years
C. 4 years
D. 5 years

159. What is the expected age at which the following milestone is first attained?

A. 4 months
B. 6 months
C. 7 months
D. 9 months

160. A 3 year old child can do all of the following except?
A. Eat with a spoon
B. Climb stairs running without support
C. Tying of shoelaces
D. Have a vocabulary of 50 to 100 words

161. A normal child has dysfluency of speech between?
A. 2 to 4 years
B. 4 to 6 years
C. 6 to 8 years
D. 8 to 10 years

162. A 3year old child is categorised as developmentally delayed if he is unable to?
A. Copy a square
B. Hop on one foot
C. Use spoon effectively without spilling
D. Catch a ball reliably

163. Bidextrous grasp develops at what age?
A. 3 months
B. 5 months
C. 7 months
D. 9 months

164. Milk is deficient in?
A. Iron and vitamin C
B. Iron and vitamin A
C. Phosphorous and vitamin C
D. Phosphorous and vitamin A

165. Breastmilk is deficient in all except?
A. Vitamin B1
B. Vitamin C
C. Vitamin D
D. Vitamin K

166. Breast milk can be stored in a refrigerator upto?
A. 4 hours
B. 8 hours
C. 12 hours
D. 24 hours

167. Compared with cow's milk, mother's milk has more?
A. Lactose
B. Vitamin D
C. Proteins
D. Fats

168. Para amino benzoic acid present in breast milk protects against infection from?
A. Plasmodium vivax
B. E coli
C. Giardia
D. Klebsiella

169. The average whey: casein ratio in breast milk is?
A. 60:40
B. 80:20
C. 20:80
D. 40:60

170. The Calcium: Phosphorous ratio in breast milk is?
A. 1:2
B. 2:1
C. 1:1
D. 3:1

171. Which among the following is a contraindication to breastfeeding?
A. Hepatitis A
B. Hepatitis B
C. CMV
D. Active untreated TB

172. Toad skin is produced due to the deficiency of?
A. Vitamin A
B. Vitamin B2
C. Vitamin D
D. Biotin

151. c	152. c	153. d	154. d	155. a	156. d	157. b	158. b	159. b	160. c	161. a
162. c	163. b	164. a	165. b	166. d	167. a	168. a	169. a	170. b	171. d	172. a

173. Casal necklace is seen due to?

A. Scurvy
B. Marasmus
C. Pellagra
D. Kwashiorkor

174. Daily protein requirement in a 2-year old child is?

A. 13 gm
B. 25 gm
C. 40 gm
D. 60 gm

175. In which stage of Vitamin A deficiency, corneal xerosis is seen?

A. Stage X1
B. Stage X2
C. Stage X3A
D. Stage X3B

176. Which gene mutation is responsible for acrodermatitis enteropathica?

A. RAS
B. SLC39A4
C. PAX6
D. SMARC45

177. Abnormalities in copper metabolism are implicated in the pathogenesis of all except?

A. Cardiomyopathy
B. Indian childhood cirrhosis
C. Menke's disease
D. Wilson disease

178. Inheritance of vitamin D dependent rickets type I?

A. Autosomal dominant
B. Autosomal recessive
C. X linked dominant
D. X linked recessive

179. Alopecia is associated with which variety of rickets?

A. X linked dominant rickets
B. Vitamin D-dependent rickets type 2
C. Nutritional rickets
D. All of the above

180. Best assessment of Vitamin C status in an infant is done by measuring levels of Vitamin C in?

A. Plasma
B. Urine
C. Buffy coat
D. Adrenal cortex

173. a	174. b	175. b	176. b	177. a	178. b	179. b	180. c

131: A. Insulin

Explanation: Fetal growth is maximally affected by insulin and insulin-like growth factors (IGFs), also called somatomedins

132: C. 12 months

Explanation: At 5 months of age, weight becomes double the birth weight and at 1 year of age, weight becomes triple the birth weight

133: B. 2 yr

Explanation: Adult height is approx. 175 to 180 cm
Half of that is 90 cm, which is attained normally at 2 years of age.

134: B. 50%

Explanation: Height at birth= 50 cm
Height at 1 year-= 75 cm
Gain in first year of life= 25 cm
Hence, % gain= 50%

135: D. 5 mg/day

Explanation: RDA for zinc in various age groups

Group	RDA for Zinc (mg/day)
Adult man	12
Adult woman: Non-pregnant	10
Adult woman: Pregnant	12
Adult woman lactating	
0 to 6 months	12
6 to 12 months	12
Children	
1-3 yr	5
4-6 yr	7
7-9 yr	8
Boys 10-12 yr	9
Girls 10-12 yr	9

136: D. 13 months

Explanation: Delayed dentition is failure of teeth to erupt by 13 months of age.

137: B. SMR 2

Explanation:

Sexual Maturity Rating Stages in Males			
SMR Stage	Pubic Hair	Penis	Testes
1	None	Preadolescent	Predolescent
2	Scanty, Long, Slightly pigmented	Minimal change / enlargement	Enlarged scrotum, pink, texture altered
3	Darker, starting to curl, small amountv	Lengthens	Larger
4	Resembles adult type, but less quantity;	Larger; glans and breath increase in size	Larger, scrotum dark
5	Adult distribution, spread to medical surface of things	Adult size	Adult size

138: A. Growth spurt

Explanation: Sequence of puberty in females
Growth spurt – Thelarche – Pubarche – Menarche
Growth spurt is the first sign of puberty
Thelarche is the first visible sign of puberty
Peak growth velocity (also known as peak growth spurt) occurs between pubarche and menarche

139: D. Thelarche

Explanation: Refer to explanation of Q no. 9

140: C. HC below -3 SD

Explanation: Microcephaly is defined as head circumference (HC) below -3 SD of the mean

141: A. Docosahexaenoic acid

Explanation: DHEA is a fatty acid exclusively present in breast milk and essential for retina and brain development.

142: A. Constitutional

Explanation: Most common cause of short stature is constitutional delay in growth

143: B. IGF-1 levels are low for chronological age

Explanation: In constitutional delay in growth, bone age is below chronological age and IGF-1 levels are low for chronological age. However, at birth, the weight and height are normal and no apparent abnormalities can be detected.

144: D. Hemorrhagic disease

Explanation: Breastmilk protects against necrotizing enterocolitis, eczema and tetany but since breast milk is deficient in Vitamin K, it increases the risk of hemorrhagic disease of newborn. Artificial top-feeds contain Vitamin K and hence they do not cause Vit K deficiency associated hemorrhagic disease.

145: D. Continue normal feeding as before

Since expected weight for age as well as height for age in the child is > 90% of expected, the child is not malnourished. Hence, routine home-based balanced diet should be continued.

More details in discussion with examples

146: C. Rate of increase of height and weight

Explanation: Rate of increase in weight and height is better than any single nutritional index

147: C. Weight for age

Explanation: Key one-liner concepts to remember:

- Marker of acute malnutrition: Weight for height
- Marker of chronic malnutrition: Height for age
- Marker of acute and chronic malnutrition: Weight for age
- Marker of acute on chronic malnutrition: Weight for height and height for age (Both)

148: A. Hepatomegaly

Explanation: Hepatomegaly and edema occur in kwashiorkor, not marasmus. All others can be seen in marasmus. Features of marasmus:

- Body weight < 60% of expected
- 6 months to 2 years
- Dry & inelastic skin
- Baggy pants appearance (AIIMS MCQ)
- Alert & irritable, buccal pad of fat in preserved till late
- Voracious appetite / old man's or monkey faces look
- Low insulin levels
- Low activity and weakness

149: A. High blood osmolarity

Explanation: Due to hypoalbuminemia, the blood osmolarity is low in Kwashiorkar. All others can be seen.

150: B. Edema and mental changes

Explanation: Kwashiorkor: 4 essential hallmark/ defining features

P - Psychomotor changes (CNS changes)
E - Edema
M - Muscle wasting
G - Growth Retardation

151: C. Gomez index

Explanation:

Age independent indicas			
Name of index	**Calculation**	**Normal value**	**Value in mainutrition**
Kanawati and McLaran's index	Mid arm circumferencs / head circumference (cm)	0.32 - 0.33	Severly malnourished <0.25
Rao and Singh's index	(weight (in kg) / height^2 (in cm)) × 100	0.14	0.12 - 0.14
Dugdale's index	weight (in kg) / height$^{1.6}$ (in cm)	0.88 - 0.97	<0.79
Quaker arm circumference measuring stick (quac stick)		Mid-arm circumference that would be expected for a given height	75-85% malnourished <75% severely malnourished
Jeliffe's ratio	Head circumference / chest circumference		Ratio <1 in a child >1 year malnourished

152: C. Vit E

Explanation: Ataxia with vitamin E deficiency (AVED) is a neurodegenerative disease belonging to the inherited cerebellar ataxias. It is mainly characterized by progressive spino-cerebellar ataxia, loss of proprioception, areflexia, and is associated with a marked deficiency in vitamin E.

153: D. 5 years

Explanation:

Developmental milestones

Language milestones

Milestone	Age of attainment
Cooing, i.e. Musical vowel sounds	3 months
Monosyllables	6 months
Bisyllables e.g. MAMA, DADA, PAPA	9 - 10 months
Speaks 4-6 words, Jargon speech	12 months
Responds to name-calling	15 months
Name familier objects, e.g. Ball	15 months
Speaks 10-15 words, Names pictures	18 months
Identifies body parts by name	18 months
Speaks simple 2-words sentences e.g. Mommy shoe	19 - 22 months
Vocabulary of 50-100 words	24 months (2years)
Sentences to combine Subject, verb, object	24 months (2years)
Uses pronouns, e.g. "I"	30 months
Known full name	30 months
Knows age and sex, Repeats sentence 6 syllables	36 months (3 years)
Tells a story, song or poem, Uses past tense	48 months (4 years)
Name 4 colours, Respeats sentence of 10 syllables	60 months (5 years)
Uses future tense	60 months (5 years)

154: D. 24 months

Explanation given in Q23.

155: A. Absence of vocalization by 6 months

Explanation: Red flag signs for development delay

Red Flag Signs in child development

Mile Stones

- No visual fixion
- No vocalization by
- No sitting without support by
- No standing alone by
- No walking alone by
- No single words by
- Lacks of imaginative play

Age

- 2 months
- 6 months
- 9 - 10 months
- 16 months
- 18 months
- 18 months
- 3 years

156: D. 36 months

Explanation: A child learns to copy a circle by 3 years and a square/rectangle by 4 years, while copying/ making a triangle comes at 5 years of age.

157: B. 3 and 4 years

Explanation: Eating with spoon without spilling comes at 18 months age onwards. Undressing under supervision comes at 3 years and knowing gender will again come at 3 years. So ans will be 3 to 4 years of age

158: B. 3 years

Explanation: A child learns to self-feed well (without spilling) between 15-18 months of age. He learns to copy a circle at 3 years and can self-dress and undress at 3 years. But independent dressing and undressing including buttons comes at 4 years of age. Hence the age of the child is 3 years.

As per Nelson, AAP

- undressing with support @2 years and Dressing with support @ 3 years
- Dressing and undressing independently (except closing buttons) 3years
- Dressing and undressing independently (including closing buttons) 4-5 years

159: B. 6 months

Explanation: The child is smiling and playing with mirror image, which is first attained at 6 months of age.

160: C. Tying of shoelaces

Explanation: Tying of shoelaces comes at 4 to 5 years of age.

161: A. 2 to 4 years

Explanation: Developmental dysfluency and stuttering commonly occur between 2 to 5 years of age according to Ghai 8th Ed.

162: C. Use spoon effectively without spilling

Explanation: Using spoon effectively to eat without spilling comes at 15 to 18 months of age according to Nelson 20th Ed. Hence if the child is unable to do so at 3 years, will be considered to be developmentally delayed. All others come after 3 years of age.

163: B. 5 months

Explanation: One-liner

164: A. Iron and vitamin C

Explanation: Milk, when used in common conversation, refers to animal milk (cow/buffalo) and animal milk is deficient in Iron and Vitamin C

165: B. Vitamin C

Explanation: Breast milk is deficient in 3 vitamins:

- Vitamin K
- Vitamin D
- Vitamin B1

Vitamin C is present in sufficient amount in breastmilk

166: D. 24 hours

Explanation: According to newbornwhocc/AIIMS guidelines

Storage of expressed breast mikl

Stored at	Can be stored for
At room temperature	6 hours
Un refrigerator	24 ho
In freezer at -4°C at -20°C	 2 weeks 3 months

167: A. Lactose

Explanation: Mother's milk has more lactose (7 g/100 ml) which protects the baby from hypoglycaemia. Protein and fat content are lower in human milk.

168: A. Plasmodium vivax

Explanation: PABA in breast milk protects against malarial infection.

169: A. 60:40

Explanation: In human milk, Whey protein is 60% and Casein protein is 40%

170: B. 2:1

Explanation: Calcium content in breast milk is 35 mg/100 ml and Phosphorous content is 15 mg/100 ml. This ratio is close to 2:1, which is ideal for optimum absorption and utilization of calcium, thereby decreasing the risk of hypocalcemic tetany.

171: D. Active untreated TB

Explanation: Although TB bacillus is not transmitted via breast milk, still it can be passed on via sputum/ respiratory secretions of the mother if she is active, untreated TB case.

172: A. Vitamin A

Explanation: Toad skin is produced due to deficiency of Vitamin A, this is known as Phrynoderma

173: C. Pellagra

Explanation: The **Casal** collar or **Casal necklace** is a clinical sign in which there is an erythematous pigmented skin rash in the distribution of a broad collar (dermatomes C3 and C4). It is seen in patients with pellagra.

174: A. 13 gm

Explanation: Daily protein requirement is around 10 gm/day for infants and 13 gm/day for 1 to 3 years of age.

175: B. Stage X2

Explanation:

World health organization re-classification of xerophthalmia signs

Classification	Ocular Signs
XN	Night blindness
XIA	Conjunctival xerosis
XIB	Bitot's spots
X2	Corneal xerosis
X3A	Corneal ulceration - keratomalacia involving one - third or less of the cornea
X3B	Corneal ulceration - keratomalacia involving one - half or less of the cornea
XS	Corneal scar
XF	Xerophthalmic fundus

176: B. SLC39A4

Explanation: As per Nelson 20th Ed, acrodermatitis enteropathica is a rare autosomal recessive disorder caused by an inability to absorb sufficient zinc from the diet. The genetic defect is in the intestinal zinc-specific transporter gene SLC39A4.

177: A. Cardiomyopathy

Explanation: Copper metabolic defects are associated with all the other three options.

178: B. Autosomal recessive

Explanation: Children with vitamin D–dependent rickets type 1, an autosomal recessive disorder, have mutations in the gene encoding renal 1α-hydroxylase, preventing conversion of 25-D into 1,25-D. These patients normally present during the 1st 2 yr of life and can have any of the classic features of rickets.

179: B. Vitamin D-dependent rickets type 2

Explanation: Patients with vitamin D–dependent rickets type 2 have mutations in the gene encoding the vitamin D receptor, preventing a normal physiologic response to 1,25-D. Levels of 1,25-D are extremely elevated in this autosomal recessive disorder. Approximately 50-70% of children have **alopecia**, which tends to be associated with a more severe form of the disease and can range from alopecia areata to alopecia totalis. Epidermal cysts are a less common manifestation.

180: C. Buffy coat

Explanation: Best assessment of Vitamin C levels in the body is done by WBC ascorbic acid estimation, for which buffy coat is usually prepared.

181. A neonate if found to have microcephaly, IUGR and multiple blueberry muffin lesions on the skin. CT brain showed calcification around the ventricles. Which among the following will be the best sample for isolation of organism?

A. Blood
B. Urine
C. Sputum
D. Biopsy from skin lesions

182. In suspected congenital CMV infection with neonate having IUGR, thrombocytopenia, periventricular calcification and hepatosplenomegaly, which is the best sample to perform PCR study?

A. Blood
B. Liver biopsy
C. Brain biopsy
D. Urine

183. In Hutchinson triad in newborn child what is not seen?

A. Interstitial keratitis
B. Notched incisors
C. Deafness
D. Cataracts

184. Which of the following is not a common manifestation of congenital rubella?

A. Deafness
B. PDA
C. Aortic stenosis
D. Cataract

185. In a neonate with limb hypoplasia, cicatricial skin lesions and IUGR, which among the following is the likely diagnosis?

A. Congenital syphilis
B. Congenital rubella
C. Congenital toxoplasmosis
D. Congenital varicella

186. What is the treatment of choice in a child born to mother who had got chicken pox in first trimester and baby found to have limb hypoplasia and scarring skin lesions?

A. No therapy
B. VZIG alone
C. Acyclovir alone
D. VZIG + Acyclovir

187. What is the therapy of choice in asymptomatic baby born to pregnant female who gets chicken pox one day prior to delivery?

A. VZIG to baby + Acyclovir to mother as well as baby
B. Only Acyclovir to mother and baby
C. VZIG to both mother and baby but no acyclovir to be given
D. Nothing active needed since child is asymptomatic

188. What is the most common long-term sequela of congenital CMV infection?

A. Intellectual dysfunction
B. Low vision due to chorioretinitis
C. Hearing loss
D. Bone marrow suppression

189. Large-sized head instead of microcephaly may be seen in which congenital infection?

A. Toxoplasmosis
B. CMV
C. Rubella
D. Syphilis

190. Which antibiotic achieves highest concentration in CSF of children suffering from congenital neurosyphilis?

A. Procaine penicillin G
B. Aqueous penicillin G
C. Benzathine penicillin G
D. All of the above

191. A child comes to OPD with oral ulcers on buccal mucosa and tongue, along with skin lesions as shown in the picture below. What is the likely causative agent?

A. Enterovirus 71
B. Coxsackievirus A16
C. Coxsackievirus A6
D. Coxsackievirus B5

192. Which among the following method is most commonly used for diagnosis of HIV infection in a 2-month old child?

A. HIV DNA PCR
B. HIV RNA PCR
C. HIV ELISA
D. P24 Ag assay

193. Most common complication of measles is?

A. Pneumonia
B. Diarrhea
C. SSPE
D. Acute otitis media

194. Most common cause of death in measles is?

A. Pneumonia
B. Diarrhea
C. SSPE
D. Acute otitis media

195. Not a feature of chicken pox rash?

A. Symmetrical
B. Mostly involves flexor surfaces
C. Centrifugal distribution
D. Pleomorphic

196. The most common site of extra-pulmonary TB in children?

A. GIT
B. CNS
C. Lymph nodes
D. Kidney

197. Which among the following is recommended in a child born to mother with active TB?

A. Stop breast feeding and start INH prophylaxis to baby
B. Continue breastfeeding and give BCG and INH prophylaxis to baby
C. Continue breastfeeding and give INH prophylaxis but avoid BCG
D. Stop breastfeeding, start INH prophylaxis and restart breastfeeding after 3 months

181. b	182. d	183. d	184. c	185. d	186. a	187. a	188. c	189. a	190. b
191. b	192. a	193. d	194. a	195. c	196. c	197. b			

198. Drug of choice for pertussis is?

A. Penicillin
B. Ceftriaxone
C. Erythromycin
D. Azithromycin

199. A child presents with bouts of cough. A characteristic inspiratory whoop is present. He is not immunised. Sample of choice for investigation is?

A. Nasopharyngeal swab
B. Tracheal aspirate
C. Cough plate culture
D. Sputum culture

200. Bull neck is seen in severe cases of which of the following?

A. Diphtheria
B. Mumps
C. Tubercular lymphadenitis
D. Goitre

201. Incorrect statement regarding post-streptococcal reactive arthritis?

A. Non-migratory arthritis
B. Good response to aspirin
C. Serological evidence of a recent group A streptococcal infection
D. Usually involves the large joints

202. Nagayama spots are seen in?

A. Measles
B. Roseola infantum
C. Mumps
D. Rubella

203. Most common childhood complication of mumps is?

A. Aseptic meningitis
B. Orchitis
C. Epididymitis
D. Pneumonia

204. Childhood infection where Steroids are indicated in severe cases:

A. Enteric fever
B. Complicated malaria
C. Pertussis
D. Infectious mononucleosis

205. Which among the following has the LEAST risk of transmission by breast milk?

A. HIV
B. CMV
C. HBV
D. HCV

206. Which among the following is the most common cause of viral myocarditis in children?

A. Coxsackie virus
B. Adeno virus
C. RSV
D. Marburg virus

207. In an unimmunized 2 year old child, there was a febrile illness, accompanied by rash, which began as erythematous macules on face and spread to the periphery. There were rose-coloured lesions in the oral cavity and lymphadenopathy involving posterior auricular and suboccipital lymph nodes. What is the likely diagnosis?

A. Mumps
B. Measles
C. Rubella
D. Chicken pox

208. First sign of infant botulism?

A. Diarrhea
B. Fever
C. Constipation
D. Cranial nerve palsies

209. Bradycardia is seen in all except?

A. Scarlet fever
B. Brucellosis
C. Lyme disease
D. Typhoid

210. In a child with acute meningococcal septicemia, what is the most common complication?

A. Osteomyelitis
B. Focal skin infarction
C. Fulminant meningitis
D. Adrenal hemorrhage

198. d	199. a	200. a	201. b	202. b	203. a	204. a	205. d	206. a	207. c
208. c	209. a	210. b							

181: B. Urine

Explanation: The features are suggestive of congenital CMV infection. Key features of congenital CMV infection include the following:

- Microcephaly, blueberry muffin spots, thrombocytopenia, hepatosplenomegaly
- "Periventricular calcification" is hallmark
- Sputum and urine samples are best for PCR study
- Viral isolation by urine culture is gold standard; Shell-vial culture/spin-enhanced culture is the technique used. Blood and sputum not reliable for isolation of the organism.
- Treatment is symptomatic. Ganciclovir in severe cases.

182: D. Urine

Explanation: Read the explanation of Q1

183: D. Cataracts

Explanation: Hutchinson triad is seen in late congenital syphilis and consists of three phenomena: interstitial keratitis, malformed teeth (**Hutchinson** incisors: notched incisors), and eighth nerve deafness.

184: C. Aortic stenosis

Explanation:

Clinical features of congenital rubella syndrome
Classic triad

- Congenital heart disease (e.g., patent ductus arteriosis, pulmonary artery stenosis, pulmonary valvular stenosis.
- Ocular defects (e.g., congenital cateracts, microphthalmos, pigmentary retinopathy, congenital glaucoma)
- Hearing loss

Congenital rubella syndrome is usually associated with a failure to thrive and developmental delay as well as as microephaly. Other common presentations at birth include:

- Purpuric rash
- Hepatosplenomegaly
- Meningoencephalitis
- Radiolucent bone
- Hepatitis
- Thrombocytopenia

185: D. Congenital varicella

Explanation: The key identifying features of congenital varicella syndrome include

- Cicatricial skin lesions
- Limb hypoplasia
- Chorioretinitis
- IUGR

186: A. No therapy

Explanation: The child has congenital varicella syndrome. Since no active varicella is present in these babies, treatment is only supportive.

187: A. VZIG to baby + Acyclovir to mother as well as baby

Explanation: Infants whose mothers demonstrate varicella in the period from 5 days prior to delivery to 2 days afterward are at high risk for severe neonatal varicella. These infants acquire the infection transplacentally as a result of maternal viremia, which may occur up to 48 hr prior to onset of maternal rash. Newborns whose mothers develop varicella during the period of 5 days before to 2 days after delivery should receive VZIG as soon as possible, even if asymptomatic. Additionally, acyclovir (10 mg/kg every 8 hr) should be given to both mother and baby.

188: C. Hearing loss

Explanation: Hearing loss is the most common long-term sequela associated with congenital CMV infection. As per Nelson 20th Ed, the failure of an infant to pass a newborn hearing screening exam should raise the possibility of congenital CMV infection. Hearing loss in the older infant and young child should also alert the clinician to the possibility of congenital CMV infection, as approximately 50% of infants with hearing loss associated with congenital CMV infection will pass an initial hearing screening exam but develop hearing loss in later infancy and early childhood.

189: A. Toxoplasmosis

Explanation: In congenital toxoplasmosis, hydrocephalus as well as microcephaly may be seen. Chorioretinitis, diffuse cerebral calcification, mental retardation also occur.

190: B. Aqueous penicillin G

Explanation: According to Nelson 20th Ed, congenital syphilis is treated with aqueous penicillin G (100,000-150,000 units/kg/24 hr divided every 12 hr IV for the 1st wk of life, and every 8 hr thereafter) or procaine penicillin G (50,000 units/kg IM once daily) given for 10 days. Both penicillin regimens are recognized as adequate therapy for congenital syphilis, but higher concentrations of penicillin are achieved in the CSF of infants treated with intravenous aqueous penicillin G than in those treated with intramuscular procaine penicillin.

191: B. Coxsackievirus A16

Explanation: Hand-foot-and-mouth disease, one of the more distinctive rash syndromes, is most frequently caused by coxsackievirus A16, sometimes in large outbreaks, and can also be caused by enterovirus 71.

192: A. HIV DNA PCR

Explanation: Diagnosis of HIV infection in young children

Test	Comment
HIV DNA PCR	Preferred test to diagnose HIV-1 subtype B infection in infants and children younger that 18 mo of age; highly sensitive and specific by 2wk of age and available; performed on peripheral blood mononuclear cells. False negatives can occur in non-B subtype HIV-1 infections
HIV culture	Expensive not easily available, required up to wk to do test; not recommended
HIV RNA PCR	Preferrded test to identify non-B subtype HIV-1 infections. Similar and specificity to HIV DNA PCR in infants and children younger than 18 mo of age, but DNA PCR is generally preferred because of greater clinical experience with that assay.

193: D. Acute otitis media

Explanation: Direct one-liner from Nelson 20th Ed

194: A. Pneumonia

Explanation: According to Nelson 20th Ed, Pneumonia is the most common cause of death in measles. It may manifest as giant cell pneumonia caused directly by the viral infection or as superimposed bacterial infection.

195: C. Centrifugal distribution

Explanation: Features of rash seen in Varicella/ Chicken pox:

- Rash appears 3-4 days after fever
- Polymorphic rash (Different types of lesions seen at the same time)
- Rash is centripetal (Centrifugal rash seen in small pox)
- Rash is common on flexural surfaces and is bilaterally symmetrical
- Rash is intensely pruritic
- May heal with scarring and pigmentary disturbances

196: C. Lymph nodes

Explanation: In children, lungs are the MC site of TB, followed by lymph nodes and GIT, respectively.

Also remember: Childhood TB is paucibacillary.

197: B. Continue breastfeeding and give BCG and INH prophylaxis to baby

Explanation: As per RNTCP guidelines and WHO recommendations, breastfeeding is not stopped in these children. We give BCG and also start INH prophylaxis @ 10 mg/kg given for 6 months. Remember that tubercle bacillus is not excreted in breast milk.

198: D. Azithromycin

Explanation: According to Nelson 20th Ed, an antimicrobial agent always is given when pertussis is suspected or confirmed, primarily to limit the spread of infection and secondarily for possible clinical benefit. Macrolides are preferred agents and Azithromycin is preferred among them for use in children.

199: A. Nasopharyngeal swab

Explanation: Culture of nasopharyngeal swab is the investigation of choice in suspected pertussis.

200: A. Diphtheria

Explanation: In diphtheria, underlying soft-tissue edema and enlarged lymph nodes can cause a bull-neck appearance. The degree of local extension correlates directly with profound prostration, bull-neck appearance, and fatality due to airway compromise or toxin-mediated complications.

201: B. Good response to aspirin

Explanation: Poststreptococcal reactive arthritis (PSRA) has been used to describe a syndrome characterized by the onset of acute arthritis following an episode of GAS pharyngitis in a patient whose illness does not fulfil the Jones criteria for the diagnosis of acute rheumatic fever. Although PSRA usually involves the large joints like the arthritis of acute rheumatic fever, it may involve small peripheral joints, as well as the axial skeleton, and is typically non-migratory, characteristics distinct from the arthritis of acute rheumatic fever. The latent period between the antecedent episode of GAS pharyngitis and PSRA may be considerably shorter (usually <10 days) than that typically seen with acute rheumatic fever (usually 14-21 days). In contrast to the arthritis of acute rheumatic fever, PSRA does not respond dramatically to therapy with aspirin or other nonsteroidal anti-inflammatory agents. In addition, PSRA is usually not migratory, and fewer patients have a fever >38°C (100.4°F). Even though no more than half of patients with PSRA who have a throat culture performed have GAS isolated, all have serologic evidence of a recent GAS infection.

202: B. Roseola infantum

Explanation: Nagayama spots consist of erythematous papules on the mucosa of the soft palate and the base of the uvula, seen in Roseola Infantum.

203: A. Aseptic meningitis

Explanation: Meningitis and meningoencephalitis are most common complications of mumps in children, whereas orchitis is most common complication in adolescents and adults.

204: A. Enteric fever

Explanation: As per Nelson 20th ed, page 1392, additional treatment with dexamethasone (3 mg/kg for the initial dose, followed by 1 mg/kg every 6 hr for 48 hr) is recommended for severely ill patients with shock, obtundation, stupor, or coma in patients with enteric fever.

205: D. HCV

Explanation: Risk of transmission via breast milk: CMV>HIV>HBV>HCV (Least)

206: A. Coxsackie virus

Explanation: Coxsackie virus is MC cause of myocarditis in children, followed by adeno virus

207: C. Rubella

Explanation: **Postnatal infection** with rubella is a mild disease not easily discernible from other viral infections, especially in children. Following an incubation period of 14-21 days, a prodrome consisting of low-grade fever, sore throat, red eyes with or without eye pain, headache, malaise, anorexia, and lymphadenopathy begins. Suboccipital, postauricular, and anterior cervical lymph nodes are most prominent. In children, the first manifestation of rubella is usually the rash, which is variable and not distinctive. It begins on the face and neck as small, irregular pink macules that coalesce, and it spreads centrifugally to involve the torso and extremities, where it tends to occur as discrete macules. About the time of onset of the rash, examination of the oropharynx may reveal tiny, rose-colored lesions (**Forchheimer spots**) or petechial hemorrhages on the soft palate. The rash fades from the face as it extends to the rest of the body so that the whole body may not be involved at any one time. The duration of the rash is generally 3 days, and it usually resolves without desquamation. Subclinical infections are common, and 25-40% of children may not have a rash.

208: C. Constipation

Explanation: **Infant botulism** differs in apparent initial symptoms of illness only because the infant cannot verbalize them. Usually, the first indication of illness is a decreased frequency or even absence of defecation, producing severe constipation.

209: A. Scarlet fever

Explanation: Relative bradycardia (temperature–pulse dissociation), when the pulse rate remains low in the presence of fever, can accompany typhoid fever, brucellosis, leptospirosis, or drug fever. Bradycardia in the presence of fever also may be a result of a conduction defect resulting from cardiac involvement with acute rheumatic fever, Lyme disease, viral myocarditis, or infective endocarditis.

210: B. Focal skin infarction

Explanation: According to Nelson 20th Ed, the most common complication of acute severe meningococcal septicaemia in children is focal skin infarction, which most commonly affects the lower limbs and can lead to substantial scarring and require skin grafting. Distal tissue necrosis in purpura fulminans may require amputation (which should be delayed to allow demarcation) in approximately 2% of survivors.

211. Which congenital heart disease has the LEAST chance of heart failure in infants?

A. TOF
B. VSD
C. TAPVC
D. ASD

212. Prevalence of congenital heart disease is?

A. 0.8%
B. 1-2%
C. 2-6%
D. 10-12%

213. Most common cardiac lesion in down's syndrome is?

A. Atrioventricular septal defect
B. VSD
C. Coarctation of aorta
D. Transposition of great vessels

214. Recurrent respiratory tract infection occur in all of the following except?

A. VSD
B. TOF
C. TGA
D. TAPVC

215. Six week old Rajesh is brought for 1st immunization with c/o feeding difficulty, forehead sweating & fast breathing. On examination he is irritable, has tachycardia, tachypnea and hepatomegaly and has not gained weight. CXR shows cardiomegaly. His mother has been told the baby has some cardiac problem at birth and was advised follow up, but she did not turn up. The most likely diagnosis is?

A. Bacterial endocarditis
B. CCF
C. Cyanotic spell
D. Cor pulmonale

216. Most common congenital heart disease is?

A. ASD
B. VSD
C. PDA
D. TOF

217. The congenital heart disease most commonly associated with pre-excitation syndrome is?

A. Double outlet RV
B. Taussig- Bing anomaly
C. Ebstein's anomaly
D. Truncus arteriosus

218. The most common congenital heart disease associated with thymic hypoplasia is?

A. PS
B. Truncus arteriosus
C. PDA
D. TOF

219. Coarctation of the aorta is common in which syndrome?

A. Down's syndrome
B. Noonan's syndrome
C. Turner's syndrome
D. Klinefelter's syndrome

220. The congenital heart disease associated with lithium intake during pregnancy?

A. TOF
B. TGA
C. ECD
D. Ebstein's anomaly

221. NADAS criteria is useful for?

A. Assessment of congenital heart disease in adults
B. Assessment of congenital heart disease in children
C. Assessment of dehydration in both children and adults
D. Assessment of congenital heart disease only in pregnancy

222. The congenital heart disease which is not ductal dependent?

A. Truncus arteriosus
B. TGA with intact ventricular septum
C. Hypoplastic left heart syndrome
D. Critical pulmonary Stenosis

223. Identify the likely disease in the child show below:

A. ASD
B. VSD
C. ECD
D. TGA

224. Which of the following features on x ray chest can differentiate an atrial septal defect (ASD) and a ventricular septal defect (VSD)?

A. Enlarged left atrium
B. Pulmonary plethora
C. Enlarged pulmonary artery
D. Enlarged aorta

225. A 2 year old child was diagnosed as a case of VSD with intermittent CCF and recurrent pneumonias. But the child was lost to follow-up and was seen 5 years later. The parents of the child reported spontaneous resolution of CCF in the child since past few weeks. What is your interpretation?

A. Parents are lying
B. Spontaneous closure of a muscular VSD
C. Restriction of L → R shunt by restrictive VSD
D. Impending eisenmenger syndrome

211. a	212. a	213. a	214. b	215. b	216. b	217. c	218. b	219. c	220. d
221. b	222. a	223. a	224. a	225. d					

226. All are true about tetralogy of fallot except?

A. Right sided aortic arch seen in 85% cases
B. Associated with CATCH – 22 syndrome
C. Persistence of a left superior vena cava is seen in
D. Multiple ventricular septal defects may be seen

227. An infant had repeated episodes of bluish discoloration of lips, which were relieved by squatting position. Chest X-ray is shown below.

What is the likely diagnosis?

A. Ostium primum ASD
B. VSD with severe pneumonia
C. TGA
D. TOF

228. All of the following are used for the treatment of cyanotic spells except –

A. Phenylephrine
B. Calcium chloride
C. Sodium bicarbonate
D. Propranolol

229. The most consistent feature suggestive of a child with TOF being in cyanotic spell is:

A. SpO2 <85%
B. Disappearance of pulmonary ejection systolic murmur
C. Syncope in the child
D. Hyperventilation

230. Eisenmenger syndrome is characterized by all except:

A. Return of left ventricle & right ventricle to normal size
B. Right ventricular hypertrophy
C. Pruning of peripheral pulmonary arteries
D. Dilatation of central pulmonary artery

231. A preterm baby with large PDA and CCF has associated renal failure. The platelet count is 50,000 & USG cranium shows IVH. The baby is also suffering form NNEC. The most appropriate modality of PDA closure would be?

A. Indomethacin 0.2 mg/kg 3 doses at the 12th hourly intervals IV/PO
B. Brufen – 10 mg/ Kg 3 doses at 24 hour interval
C. Bedside ligation of PDA
D. PGE, infusion

232. True about 1 year old child with PDA is?

A. Symptoms similar to aorto pulmonary window
B. Chances of spontaneous closure high
C. Indomethacin may help in closure
D. Endocarditis is rare

233. Congestive heart failure in children Is first diagnosed by:

A. Tachycardia and tender hepatomegaly
B. Increased JVP
C. Increased JVP+ pedal edema
D. Hypotension

234. The only cyanotic disease showing LV hypertrophy on CXR is:

A. Tricuspid atresia
B. Ebstein's anomaly
C. Truncus arteriosus
D. PDA with Eisenmenger

235. Identify the disease associated with this CXR:

A. TOF
B. TGA
C. TAPVC
D. Truncus arteriosus

236. RIB notching of 4-9 th ribs is seen in?

A. Aortic aneurysm
B. Aortic dissection
C. Coarctation of aorta
D. Diaphragmatic hernia

237. A cyanotic newborn is suspected of having CHD. She has a left ventricular impulse, with holosystolic murmur, audible at left sternal border with single S_2. CXR shows decreased pulmonary blood flow, oligemic lung fields. ECG shows left axis deviation. The most probable diagnosis is?

A. TOF
B. TGA
C. Tricuspid atresia
D. TAPVC

238. A child with TOF presents with mild fever, vomiting and headache for the past 3 days and is rushed to the casualty early morning in altered sensorium. On examination he is cyanosed, has clubbing, ESM murmur is heard in pulmonary area. He throws convulsions in ER and neurological examination shows paucity of movements and Babinski's on R side. The most probable diagnosis?

A. Paradoxical emboli to CNS
B. Cyanotic spell
C. Brain abscess
D. Meningitis

226. a	227. d	228. b	229. b	230. a	231. c	232. a	233. a	234. a	235. c
236. c	237. c	238. c							

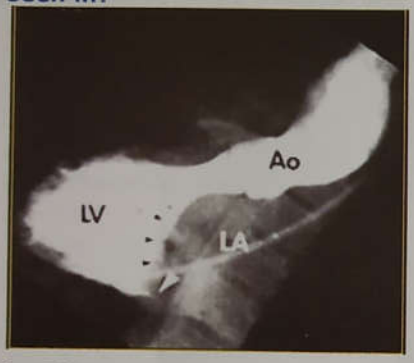
239. Potts shunt is anastomosis of:

A. Right subclavian artery to right pulmonary artery
B. Descending aorta to left pulmonary artery
C. Left subclavian to left pulmonary artery
D. Ascending aorta to right pulmonary

240. Right sided aortic arch is commonly associated with?

A. Truncus arteriosus
B. TAPVC
C. Endocardial cushion defect
D. PDA

241. The following visualisation on angiography is seen in?



A. ECD
B. VSD
C. Truncus arteriosus
D. Preductal CoA

242. The following CXR appearance is seen in which condition?

A. TOF
B. TGA
C. TAPVC
D. Truncus arteriosus

243. Box-shaped heart is seen in?

A. TOF
B. TGA
C. Ebstein's anomaly
D. Tricuspid atresia

244. Drug used to keep DA open is?

A. PGE1
B. PGE2
C. PGI2
D. TXA2

245. Which congenital heart disease worsens on starting PGE1 infusion?

A. TGA with intact ventricular septum
B. Obstructive TAPVC
C. Ebstein's anomaly
D. Pulmonary atresia

246. Most common cause of hypertension in children?

A. Renovascular disease
B. Essential hypertension
C. Coarctation of aorta
D. Pheochromocytoma

247. Mechanism of acute rheumatic fever is?

A. Immune complex disease
B. Bacterial toxin mediated injury
C. Molecular mimicry
D. All of the above

248. Duration of prednisolone in carditis in acute rheumatic fever is?

A. 2 weeks
B. 6 weeks
C. 12 weeks
D. 24 weeks

249. Not a major criteria for diagnosis of acute rheumatic fever in high-risk populations according to latest modified jones criteria?

A. Monoarthritis
B. Polyarthralgia
C. Subcutaneous nodules
D. Elevated CRP >3 mg/dl

250. Which class of drugs is used as initial empirical therapy of childhood CCF?

A. Diuretics
B. CCBs
C. ACE inhibitors
D. ARBs

239. b	240. a	241. a	242. b	243. c	244. a	245. b	246. a	247. c	248. c
249. d	250. a								

211: A. TOF

Explanation: TOF carries least risk of CCF, followed by ASD.

212: A. 0.8%

Explanation: According to Nelson 20th Ed, congenital heart disease occurs in approximately 0.8% of live births. The incidence is higher in stillborns (3-4%), spontaneous abortuses (10-25%), and premature infants (approximately 2% excluding patent ductus arteriosus [PDA]).

213: A. Atrioventricular septal defect

Explanation: The most common cardiac disease in down syndrome is Endocardial cushion defect, also known as atrioventricular septal/atrioventricular canal defect.

214: B. TOF

Explanation: TOF is a condition with decreased pulmonary blood flow and thus not prone to recuurent respiratory tract infections.

215: B. CCF

Explanation: The key clues to underlying CCF include:

- Failure to gain weight
- Recurrent respiratory infections or pneumonias
- Feeding difficulties and sweating on forehead while feeding
- Tachycardia
- Hepatomegaly
- Cardiomegaly
- Basal crepitations
- Pedal edema

The child in question has most of these, suggestive of CCF.

216: B. VSD

Explanation: Frequency of all congenital heart diseases

Lesion	% of all Leasions
Ventricular septal defect	35-30
Atrial septal defect (secundum)	6-8
Patent ductus arterious	6-8
Coarctation of aorta	5-7
Tetralogy of Fallot	5-7
Pulmonary valve stenosis	5-7
Aortic valve stenosis	4-7
D-Transposition of great arteries	3-5
Hypoplastic left ventricle	1-3
Hypoplastic right ventricle	1-3
Truncus arterious	1-2
Total anomalous pulmonary venous return	1-2
Tricuspid atresia	1-2
Single ventricle	1-2
Double outlet right ventricle	1-2
Others	5-10

217: C. Ebstein's anomaly

Explanation: Ebstein's anomaly is frequently associated with Wolff- Parkinson- White syndrome (WPW syndrome), which is a type of pre-excitation syndrome.

218: B. Truncus arteriosus

Explanation: MC congenital heart disease in thymic hypoplasia/DiGeorge syndrome : Truncus arteriosus> TOF

219: C. Turner's syndrome

Explanation:

Syndrome	Features
Down syndrome	Endocardial cushion defect, VSD, ASD
18 Trisomy	VSD, ASD, PDA, Coarctation of aorta, bucuspid aortic / pulmonary value
Turner syndrome	Bicuspid aortic valve, coarctation of india
Charge Syndrome	VSD, ASD, PDA, TOG, Endocardial cushion defect
Vater Syndrome	VSD, TOF, ASD, PDA

220: D. Ebstein's anomaly

Explanation: Ebstein's anomaly is a rare congenital heart disease characterised by downward displacement of the septal and posterior leaflets of tricuspid valve into RV cavity. It is often associated with maternal Li ingestion in pregnancy.

221: B. Assessment of congenital heart disease in children

Explanation: **NADA'S CRITERIA:** PRESENCE OF ONE MAJOR OR TWO MINOR FULFILLS CRITERIA. It indicates underlying congenital heart disease in children

Major

1) Systolic murmur gr III or more associated with thrill
2) Diastolic murmur
3) Cyanosis
4) CHF

Minor

1) Systolic murmur < gr III
2) Abnormal S2
3) Abnormal ECG
4) Abnormal Xray
5) Abnormal BP

222: A. Truncus arteriosus

Explanation: Ductus dependent congenital heart diseases

Left-Sided Obstruction (Systemic Circulation Impaired)	Right-Sided Obstruction (Pulmonary Circulation Impaired)
Hypoplastic left heart syndrome	Tetralogy of fallot with pulmonary atresia
Critical aortic stenosis	Pulmonary artersia
Coarctation of the aorta	Critical pulmonary stenosis
Interruted aortic arch	Tricuspid atresia
	Severe Ebstein's anomaly
	Complete transposition of the great arteries with intact interventricular septum

TGA with IVS is sometimes sub-classified as ductus dependent oxygenation.

In Truncus arteriosus, there is mixing of oxygenated and deoxygenated blood and is not dependent on open ductus arteriosus for survival.

223: A. ASD

Explanation: The child has limb defects, suggestive of Holt-Oram syndrome. It is commonly associated with atrial septal defects (ASD).

224: A. Enlarged left atrium

Explanation: Left atrium is often enlarged in VSD, whereas it is never enlarged in ASD and can be useful to distinguish the two conditions.

225: D. Impending eisenmenger syndrome

Explanation: Spontaneous resolution of CCF symptoms in a child with VSD suggests 2 possibilities:

- Either it was a muscular VSD, which closed spontaneously (Common upto 2 yr age)
- Or it is turning into severe pulmonary hypertension. When pulm HTN starts increasing, pulmonary blood flow temporarily falls and features of CCF may improve but it is a dangerous thing to occur since the child will soon develop reversal of shunt if immediate surgery is not performed. Such a condition is known as impending Eisenmenger syndrome, where frank reversal of sunt has still not happened but severe pulm HTN is evident clinically as well as on ECHO.

Since this child is presenting late, chance of spontaneous closure of muscular VSD is low and the likely cause is impending Eisenmenger syndrome.

226: A. Right sided aortic arch seen in 85% cases

Explanation: According to Nelson 20th Ed, a right aortic arch occurs in about 20% of patients, and other anomalies of the pulmonary arteries and aortic arch may also be seen. Persistence of a left superior vena cava draining into the coronary sinus is common but not a concern. Multiple VSDs are occasionally present and must be diagnosed before corrective surgery. Tetralogy of Fallot can be associated with **DiGeorge syndrome** or **Shprintzen velocardiofacial syndrome**, also known by the acronym **CATCH 22** (cardiac defects, abnormal facies, thymic hypoplasia, cleft palate, hypocalcemia).

227: D. TOF

Explanation: History of cyanosis and relief on squatting is typically seen in patients with tetralogy of fallot (TOF). The typical radiologic configuration as seen in the anteroposterior view consists of a narrow base, concavity of the left heart border in the area usually occupied by the pulmonary artery, and normal overall heart size. The hypertrophied right ventricle causes the rounded apical shadow to be uptilted so that it is situated higher above the diaphragm than normal and pointing horizontally to the left chest wall. The cardiac silhouette has been likened to that of a boot ("coeur en sabot"). The hilar areas and lung fields are relatively clear because of diminished pulmonary blood flow or the small size of the pulmonary arteries, or both.

228: B. Calcium chloride

Explanation: There is no role of Calcium chloride in management of cyanotic spell in TOF. Management strategies include:

- Knee-chest position
- Humidified O2
- IV sodium bicarbonate 1 meq/kg
- SC Morphine 0.1 mg/kg
- Propranolol/metoprolol
- Phenylephrine
- Severe cases: Ketamine, Intubation and ventilation
- Emergency palliative surgery in refractory cases

229: B. Disappearance of pulmonary ejection systolic murmur

Explanation: Cyanotic spell or tet spell is a common complication of TOF in young children. It is characterised by worsening of cyanosis, hyperventilation and absence of any murmur on auscultation. Sudden disappearance of previously heard pulmonary ejection systolic murmur is a sure sign that child is in cyanotic spell.

230: A. Return of left ventricle & right ventricle to normal size

Explanation: Roentgenographically, the heart varies in size from normal to greatly enlarged; the latter usually occurs late in the course of the disease. The main pulmonary artery is generally prominent. The pulmonary vessels are enlarged in the hilar areas and taper rapidly in caliber in the peripheral branches. The right ventricle and atrium are prominent. The electrocardiogram shows marked right ventricular hypertrophy. The ventricles do not return to normal size.

231: C. Bedside ligation of PDA

Explanation: Since child has NEC, thrombocytopenia and IVH, medical closure of PDA is contraindicated and hence surgical ligation is recommended.

232: A. Symptoms similar to aorto pulmonary window

Explanation: An aortopulmonary window defect consists of a communication between the ascending aorta and the main pulmonary artery. It mimics presentation of PDA. Remember that any PDA beyond 3 months of age doe not undergo spontaneous closure and needs surgical ligation since it poorly responds to medical therapy.

233: A. Tachycardia and tender hepatomegaly

Explanation: Hepatomegaly is the most consistent sign of CCF in children.

234: A. Tricuspid atresia

Explanation: In tricuspid atresia, no outlet from the right atrium to the right ventricle is present; the entire systemic venous return leaves the right atrium and enters the left side of the heart by means of the foramen ovale or, most often, through an atrial septal defect or a ventricular septal defect. The combination of cyanosis and left axis deviation on the electrocardiogram is highly suggestive of tricuspid atresia.

235: C. TAPVC

Explanation: This is snowman/figure of 8 appearance seen in supracardiac TAPVC

236: C. Coarctation of aorta

Explanation: **Notching** of the **inferior border of the ribs, mainly 3rd to 8th ribs** from pressure erosion by enlarged collateral vessels is common by late childhood in coarctation of aorta.

237: C. Tricuspid atresia

Explanation: LV hypertrophy with cyanosis in neonatal period is important clue for Tricuspid atresia.

238: C. Brain abscess

Explanation: Febrile illness with seizures and focal neurological deficits in a child with TOF are suggestive of brain abscess. Remember that paradoxical emboli will not have fever or preceding symptoms for 3 days.

239: B. Descending aorta to left pulmonary artery

Explanation: Palliative shunt surgeries in TOF:

- Pott's shunt: Left PA is joined to descending aorta
- Waterston's shunt: Right PA is joined to ascending aorta
- Modified Blalaock-Taussig Shunt: Subclavian artery is joined to pulmonary artery

240: A. Truncus arteriosus

Explanation: Explanation: Right sided aortic arch is most commonly associated with Truncus arteriosus, followed by TOF

241: A. ECD

Explanation: This is goose-neck deformity seen on angiography in endocardial cushion defect.

242: B. TGA

Explanation: This is egg-on-side appearance seen in D-TGA.

243: C. Ebstein's anomaly

Explanation: A one-liner.

244: A. PGE1

Explanation: Synthetic PGE1, known as Alprostadil is used in various complex ductus dependent heart diseases to keep the ductus arteriosus open till surgery can be performed.

245: B. TGA with intact ventricular septum

Explanation: Obstructive TPAVC is characterised by severe PAH and low pulmonary blood flow. By giving PGE1, ductus arteriosus becomes open and pulmonary blood is further diverted into the systemic circulation, leading to fall in oxygenation and worsening of underlying cyanosis. All other options are ductus dependent lesions which benefit from PGE1 infusion.

246: A. Renovascular disease

Explanation: Renovascular causes are the most common cause of hypertension in children.

247: C. Molecular mimicry

Explanation: The antigenicity of several Group A beta-hemolytic streptococcus cellular and extracellular epitopes and their immunologic cross-reactivity with cardiac antigenic epitopes lends support to the hypothesis of molecular mimicry. Common epitopes are shared between certain GAS components (e.g., M protein, cell membrane, group A cell wall carbohydrate, capsular hyaluronate) and specific mammalian tissues (e.g., heart valve, sarcomere, brain, joint). This causes damage to native tissue by immune system.

248: C. 12 weeks

Explanation: Steroids in acute rheumatic fever-associated carditis

Moderate to severe carditis Steroids

Regime I

Prednisolone: 2mg/kg/d, maximum 80mg/day till ESR normalized usually 2 weeks. Taper over 2-4 weeks, reduce dose by 2.5 - 5mg every 3rd day.

Star aspirin 50 - 75mg/kg/d simultaneously, to complete total 12 weeks (Level of evidence: Class I)

Regime II

Prednisolone same doses × 3-4 weeks.

Taper slowly to cover total period of 10 - 12 weeks (Level of evidence - Class IIb)

249: D. Elevated CRP >3 mg/dl

Explanation: **Latest modified Revised jones criteria**

MAJOR CRITERIA	MAJOR CRITERIA
Low-risk populations	Moderate- and high-risk populations
Carditis	**Carditis**
• Clinical and/or subclinical	• Clinical and/or subclinical
Arthritis • Polyarthritis only	**Arthritis** • Monoarthritis or polyarthritis • Polyarthralgia
Chorea	Chorea
Erythema marginatum	Erythema marginatum
Subcutaneous nodules	Subcutaneous nodules
MINOR CRITERIA	MINOR CRITERIA
Low-risk populations	Moderate- and high-risk populations
Polyarthralgia Fever (≥38.5°C) ESR ≥60 mm in the first hour and/or CRP ≥3.0 mg/dL Prolonged PR interval, after accounting for age variability (unless carditis is a major criterion)	Monoarthralgia Fever (≥38°C) ESR ≥30 mm/h and/or CRP ≥3.0 mg/dL Prolonged PR interval, after accounting for age variability (unless carditis is a major criterion)

250: A. Diuretics

Explanation: According to Nelson 20th Ed, diuretics are usually the first mode of therapy initiated in patients with congestive heart failure in childhood. **Furosemide** is the most commonly used diuretic in Paediatric patients with heart failure.

251. Which of the following statement is false about low sacral meningomyelocele?

A. Spasticity and weakness of the lower limbs is seen
B. Hydrocephalus is seen
C. Bladder incontinence may be seen
D. Bowel incontinence is present

252. All of the following are neural tube defects except?

A. Meningomyelocele
B. Anencephaly
C. Holoprosencephaly
D. Encephalocele

253. In a lady with previous history of child with neural tube defects, what amount of folic acid (in microgram) is to be given as prophylaxis pre-conceptionally?

A. 40
B. 400
C. 500
D. 4000

254. Banana-shaped sign on antenatal USG is seen in?

A. Aicardi syndrome
B. Dandy walker syndrome
C. Arnold chiari malformation
D. Holoprosencephaly

255. The inheritance pattern of neural tube defects is?

A. Autosomal dominant
B. Multifactorial
C. Mitochondrial
D. Autosomal recessive

256. Content of meningocele?

A. Duramater
B. Spinal cord
C. Brain tissue
D. Cauda equina

257. Meningomyelocele most commonly involves which region of spine?

A. Cervico-dorsal
B. Dorso-lumbar
C. Lumbo-sacral
D. Sacro-cocyygeal

258. A case of meningomyelocele was posted for surgery. Till surgery is done, the covering of the sac should be covered with a gauze soaked in?

A. Normal saline
B. Tincture iodine
C. Methylene blue
D. Povidone iodine

259. A preterm baby was delivered with the following anomaly. What is the likely diagnosis?

A. Porencephaly
B. Meningomyelocele
C. Encephalocele
D. Anencephaly

260. A 7-day-old infant has a leaky meningomyelocele. The most useful test for diagnosis and management is?

A. Local wound swab culture
B. Urine culture
C. Rectal swab culture
D. Blood culture

261. A one-year-old child was brought to Paediatric neurology OPD with the complaints of frequent generalised seizures and failure to thrive. On examination, child had severe developmental delay with microcephaly. His MRI brain is shown below. What is the likely diagnosis?

A. Schizencephaly
B. Holoprosencephaly
C. Polymicrogyria
D. Lissencephaly

262. A child has mental retardation, refractory seizures, hemivertebrae and absence of corpus callosum. What is the likely diagnosis?

A. Moebius syndrome
B. Miller-Dicker syndrome
C. Aicardi syndrome
D. Joubert syndrome

251. a	252. c	253. d	254. a	255. b	256. a	257. c	258. a	259. d	260. d
261. d	262. c								

263. **An infant born to mother taking Valproate in pregnancy was suspected to have spina bifida occulta. Which among the following is NOT an indication for neuroimaging in the child?**
A. Dermal sinus
B. Hyperpigmented patch
C. Tuft of hair
D. Coccygeal pit

264. **Increased risk of deformational plagiocephaly is seen with?**
A. Female child
B. High birth order
C. Development delay
D. Exclusive breast feeding

265. **Most common cause of hydrocephalus in children is?**
A. Post-inflammatory obstruction
B. Budd-Chiari syndrome
C. Brain tumour
D. Perinatal injury

266. **Not seen in Dandy-Walker malformation?**
A. Cerebellar vermis hypoplasia
B. Fourth ventricle dilatation
C. Narrow posterior fossa
D. Hydrocephalus

267. **Most common cause of obstructive hydrocephalus is?**
A. Brain tumours
B. Aqueductal stenosis
C. Tubercular meningitis
D. Pneumococcal meningitis

268. **Which among the following is the fastest way to decrease raised ICP in a child who is having unequal pupils and signs of impending herniation?**
A. I.V. Mannitol
B. I.V. Hypertonic saline (3%)
C. Hyperventilation
D. I.V. Frusemide

269. **All are signs of hydrocephalus in a neonate except?**
A. Enlarged head
B. Sunset sign
C. Crack pot sign
D. Bulging fontanelle

270. **Surgical procedure of choice in a child with hydrocephalus?**
A. Endoscopic ventriculostomy
B. VP shunt
C. TIPSS
D. External ventricular drainage

271. **Brachycephaly is due to fusion of?**
A. Parietal suture
B. Sagittal suture
C. Lambdoid suture
D. Coronal suture

272. **Most common cause of neonatal seizures?**
A. HIE
B. Intracranial HGE
C. CNS infection
D. Metabolic disease.

273. **Drug of choice for neonatal seizures is**
A. Phenytoin
B. Phenobarbitone
C. Diazepam
D. Sodium valproate

274. **Most common site of focus for mesial temporal sclerosis in adolescents is?**
A. Amygdala
B. Hippocampus
C. Basal ganglia
D. Cerebellum

275. **Burst suppression pattern on EEG is seen in:**
A. HSV encephalitis
B. Prion disease
C. Anoxic brain damage
D. Petit mal epilepsy

276. **A child started on AEDs for complex partial seizures developed severe mucocutaneous rash and ulcers involving mucous membrane. What could be the likely agent responsible?**
A. Lamotrigine
B. Carbamazepine
C. Valproate
D. Zonisamide

277. **The risk for future epilepsy if a child with febrile seizures has neurodevelopmental anomalies is**
A. 1%
B. 15%
C. 22%
D. 33%

278. **All of the following are used for managing Paediatric status epilepticus except?**
A. Phenytoin
B. Diazepam
C. Thiopentone sodium
D. Carbamazepine

279. **Drug of choice in Landau-Kleffner syndrome?**
A. Valproate
B. Phenytoin
C. Rufinamide
D. Carbamazepine

280. **Most common cause of obstructive hydrocephalus is?**
A. Brain tumours
B. Aqueductal stenosis
C. Tubercular meningitis
D. Pneumococcal meningitis

281. **In a child with pneumococcal meningitis, duration of therapy for intravenous antibiotic is?**
A. 5-7 days
B. 7-10 days
C. 10-14 days
D. 14-21 days

282. **A 12-year-old girl has recent-onset dropping of objects on hand on getting up in the morning. Her elder sister, who is 16 years old has similar symptoms since last 2 years and EEG done showed a characteristic pattern. What is the likely diagnosis?**
A. Choreoathetoid epilepsy
B. Jacksonian march
C. Juvenile myoclonic epilepsy
D. Rolandic epilepsy

283. **A 3-year-old child with acute onset of fever of 104°F developed febrile seizure and was treated. To avoid future recurrence of seizure attack, what should be given?**
A. Paracetamol+ Diazepam
B. Oral diazepam 6th hourly
C. Paracetamol 400 mg 6th hourly
D. IV diazepam infusion over 12 hours

263. d	264. c	265. c	266. c	267. b	268. c	269. c	270. b	271. d	272. a	273. b
274. b	275. c	276. b	277. d	278. d	279. a	280. b	281. c	282. c	283. b	

284. All of the following drugs are used for absence seizures except?

A. Clonazepam
B. Phenytoin
C. Valproate
D. Ethosuximide

285. Absence seizures are characterized in EEG by?

A. 3 Hz spike and wave
B. 2 Hz spike and wave
C. Brief bursts of polyspikes
D. Hypsarrythmia

286. All are features of absence seizures except?

A. Usually seen in childhood
B. 3 Hz spike wave seen in EEG
C. Post-ictal confusion
D. Precipitation by hyperventilation

287. Drug of choice in infantile spasm?

A. ACTH
B. Vigabatrin
C. Carbamazepine
D. Ethosuximide

288. Drug of choice in infantile spasm in tuberous sclerosis?

A. ACTH
B. Vigabatrin
C. Carbamazepine
D. Ethosuximide

289. DOC in simple partial seizure in children is?

A. Phenytoin
B. Valproic acid
C. Carbamazepine
D. Phenobarbitone

290. Automatisms are seen in?

A. Complex partial seizure
B. Simple partial seizures
C. Infantile spasms
D. Absence seizures

291. Not a cause of meningitis in a 12-year-old child?

A. Listeria
B, Strep pneumoniae
C. Pneumococcus
D. Staph aureus

292. A child has CSF counts of 150 cells/mm3, CSF protein of 100 mg% and CSF glucose of 75 mg%. What is the likely diagnosis?

A. Tubercular meningitis
B. Pyogenic meningitis
C. Fungal meningitis
D. Aseptic meningitis

293. In Paediatric brain abscess, intravenous antibiotics are given for a total duration of?

A. 7-10 days
B. 2-3 weeks
C. 4-6 weeks
D. 6-12 weeks

294. Highest mortality in childhood bacterial meningitis is seen with?

A. Pneumococcus
B. E. coli
C. Neisseria meningitides
D. H. influenzae

295. Most common cause of bacterial meningitis in a child with CSF rhinorrhea?

A. Streptococcus agalactiae
B. Streptococcus pneumoniae
C. Escherichia coli
D. Staphylococcus aureus

296. Bulbar palsies and intellectual dysfunction is commonly seen in which type of cerebral palsy?

A. Spastic diplegic CP
B. Spastic quadriplegic CP
C. Spastic hemiplegic CP
D. Choreoathetoid CP

297. Prior to discharge of a patient of H. influenza meningitis the essential investigation to be done is?

A. EEG
B. BERA
C. Developmentscreening test
D. ECG

298. Not true regarding Myotonic dystrophy?

A. Distal muscles are affected earlier than proximal muscles
B. Cardiac and endocrine abnormalities are common
C. Severity of myotonia is determined by degree of weakness in muscles
D. Immune abnormalities with low serum IgG is commonly seen

299. A boy presented with weakness in lower limbs, calf hypertrophy, positive gower's sign and an elevated CPK value of 10,000. The most likely diagnosis is?

A. Duchenne muscular dystrophy
B. Spinal Muscular atrophy
C. Myotonia congenital
D. Myotonic dystrophy

300. Drisapersen is a new drug effective in which among the following Paediatric disorders?

A. Myasthenia gravis
B. Guillain barre syndrome
C. Friedrich ataxia
D. Duchenne's muscular dystrophy

284. b	285. a	286. c	287. a	288. b	289. c	290. a	291. a	292. d	293. c
294. a	295. b	296. b	297. b	298. c	299. a	300. d			

251: A. Spasticity and weakness of the lower limbs is seen

Explanation: According to Nelson 20th Ed, a lesion in the low sacral region causes bowel and bladder incontinence associated with anesthesia in the perineal area but with no impairment of motor function. Hydrocephalus is often associated with meningomyelocele.

252: C. Holoprosencephaly

Explanation: Majordefectsincluded in neural tubedefects (NTDs)

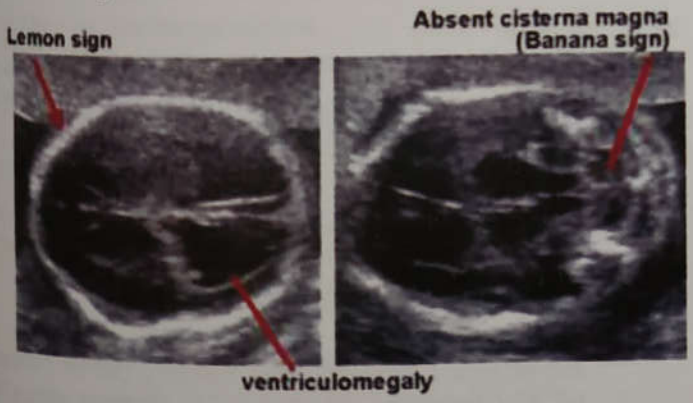
- Spinabifidaocculta
- Meningocele
- Myelomeningocele
- Encephalocele
- Anencephaly
- Dermal sinus
- Tethered cord
- Syringomyelia
- Diastematomyelia
- Lipoma

253: D. 4000

Explanation: Recommendation is to give 0.4 mg (400 microgram) in all pregnant females, whereas in high-risk women with prior NTD history, we give 4 mg (4000 microgram).

254: A. Aicardi syndrome

Explanation: Banana sign on antenatal USG is seen in Arnold chiari malformation due to absent cisterna magna.



255: B. Multifactorial

Explanation: Although the precise cause of NTDs remains unknown, evidence suggests that many factors, including hyperthermia, drugs (valproic acid), malnutrition, low red cell folate levels, chemicals, maternal obesity or diabetes, and genetic determinants (mutations in folate-responsive or folate-dependent enzyme pathways) can adversely affect normal development of the CNS from the time of conception. In some cases, an abnormal maternal nutritional state or exposure to radiation before conception increases the likelihood of a congenital CNS malformation.

256: A. Duramater

Explanation: A meningocele is formed when the meninges herniate through a defect in the posterior vertebral arches or the anterior sacrum. The spinal cord is usually normal and assumes a normal position in the spinal canal.

257: C. Lumbo-sacral

Explanation: According to Nelson 20th Ed, a myelomeningocele may be located anywhere along the neuraxis, but the lumbosacral region accounts for at least 75% of the cases.

258: A. Normal saline

Explanation: Covering with saline soaked gauze keeps it hydrated and prevents from external trauma and infection. Other substances mentioned can be irritant.

259: D. Anencephaly

Explanation: This is the classic appearance of a child with anencephaly. An anencephalic infant presents a distinctive appearance with a large defect of the calvarium, meninges, and scalp associated with a rudimentary brain, which results from failure of closure of the rostral neuropore, the opening of the anterior neural tube.

260: D. Blood culture

Explanation: In any leaky MMC, CSF culture and blood culture are recommended to detect and treat infections.

261: D. Lissencephaly

Explanation: The child is having features and MRI suggestive of Lissencephaly. Note the absence of cerebral sulci and the maldeveloped sylvian fissures associated with enlarged ventricles. Lissencephaly, or agyria, is a rare disorder that is characterized by the absence of cerebral convolutions and a poorly formed sylvian fissure, giving the appearance of a 3-4 mo fetal brain. The condition is probably a result of faulty neuroblast migration during early embryonic life and is usually associated with enlarged lateral ventricles and heterotopias in the white matter. These infants present with failure to thrive, microcephaly, marked developmental delay, and a severe seizure disorder. Ocular abnormalities are common, including hypoplasia of the optic nerve and microphthalmia. Lissencephaly can occur as an isolated finding, but it is associated with Miller-Dieker syndrome in approximately 15% of cases.

262: C. Aicardi syndrome

Explanation: **Aicardi syndrome** represents a complex disorder that affects many systems and is typically associated with agenesis of the corpus callosum, distinctive chorio-retinal lacunae, and infantile spasms. Patients are almost all female, suggesting a genetic abnormality of the X chromosome (it may be lethal in males during fetal life). Seizures become evident during the 1st few

mo and are typically resistant to anticonvulsants. An electroencephalogram shows independent activity recorded from both hemispheres as a result of the absent corpus callosum and shows often hemihypsarrhythmia. All patients have severe intellectual disability and can have abnormal vertebrae that may be fused or only partially developed (hemivertebra). Abnormalities of the retina, including circumscribed pits or lacunae and coloboma of the optic disc, are the most characteristic findings of Aicardi syndrome.

263: D. Coccygeal pit

Explanation: Indications for neuroimaging in spina bifida/spinal dysraphism:

Imaging Indicated
Subcutaneous mass of lipona
Hairy patch
Dermal sinus
Atypical dimples (deep, >5mm, >25mm from anal verge)
Vascular lesion, e.g., hemangioma or telagiectasia
Skin appendages or polypoid lesions, e.g., skin tags, tail-like appendages
Scar-Like lesions
Imaging Uncertain
Hyperpigmented patches
Deviation of the gluteal fold
Imaging Not Required
Simple dimples (<5mm, <25mm from anal verge)
Coccygeal pits

264: C. Development delay

Explanation: Deformational plagiocephaly (DP), also known as positional plagiocephaly, is the development of cranial flattening and asymmetry in the infant as a result of extrinsic molding forces placed on the skull, such as consistently sleeping on the same area of the head.

Factors that increase risk for Deformational Plagiocephaly
Male
Firstborn child
Limited passive neck rotation at birth (e.g., congenital torticollis)
Developmental delay
Sleep position is supine at birth and at 6 weeks
Bottle feeding only
Tummy time <3 times/day
Lower activity level, slower milestone achievement
Sleeping with head to same side, positional preference

265: A. Post-inflammatory obstruction

Explanation: As a group, inflammatory causes are most common.

266: C. Narrow posterior fossa

Explanation: **Dandy-Walker malformation** is part of a continuum of posterior fossa anomalies that include cystic dilation of the fourth ventricle, hypoplasia of the cerebellar vermis, hydrocephalus, and an enlarged posterior fossa with elevation of the lateral venous sinuses and the tentorium. Extracranial anomalies are also seen. Variable degrees of neurologic impairment are usually present.

267: B. Aqueductal stenosis

Explanation: Causes of hydrocephalus

- Obstructive hydrocephalus: Congenital aqueductal stenosis
- Non-obstructive hydrocephalus: Subarachnoid haemorrhage in western countries, TB meningitis in India

268: C. Hyperventilation

Explanation: Hyperventilation is the fastest way to decrease raised ICP in acute emergency setting. Acts within 3 to 5 min. Mannitol bolus is second fastest way.

269: C. Crack pot sign

Explanation: Although all four are the features of hydrocephalus, however, MacEwan's cracked pot sign is seen late and virtually never in the neonatal period.

270: B. VP shunt

Explanation: Most cases of hydrocephalus require extracranial shunts, particularly a ventriculoperitoneal shunt.

271: D. Coronal suture

Explanation: Fusion of both *coronal* sutures leads to a head shape called "brachycephaly." This causes restriction of growth of the anterior fossa resulting in a shorter and wider than normal skull.

272: A. HIE

Explanation: Anoxic damage leading to hypoxic ischemic encephalopathy (HIE) is the overall MC cause of neonatal seizures.

273: B. Phenobarbitone

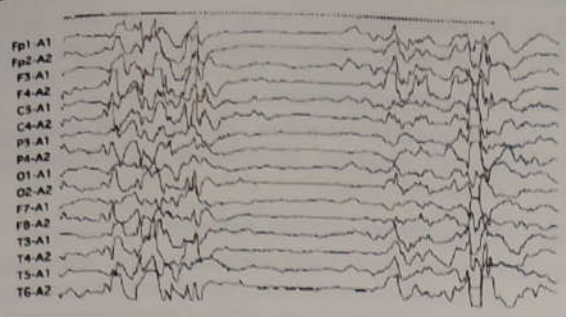
Explanation: Phenobarbitone is the drug of choice in neonatal seizures. The dose is 20 mg/kg/IV slowly over 20 minutes (not faster than 1 mg/kg/min).

274: B. Hippocampus

Explanation: Temporal lobe epilepsy can be caused by any temporal lobe lesion. A common cause is **mesial** (also termed **medial**) **temporal sclerosis**, a condition often preceded by febrile seizures and, rarely, genetic in origin. Pathologically, these patients have atrophy and gliosis of the hippocampus and, in some, of the amygdala. It is the most common cause of surgically remediable partial epilepsy in adolescents and adults.

275: C. Anoxic brain damage

Explanation: BURST SUPPRESSION PATTERN is seen in Anoxic brain damage.



276: B. Carbamazepine

Explanation: Both lamotrigine and carbamazepine can lead to severe mucosal lesions and rash, producing toxic epidermal necrolysis/Stevens Johnson syndrome. However, Carbamazepine is more commonly used in children for partial epilepsy and has slightly higher risk when started in young children to produce such effects.

277: D. 33%

Explanation: Risk factors for future epilepsy in febrile seizures

Risk Factor	Risk for Subsequent Epilepsy
Simple febrile seizure	1%
Recurrent febrile seizures	4%
Complex febrile seizures (more than 15 min duration of recurrent within 24hr)	6%
Fever <1 hr before febrile seizure	11%
Family history of epilepsy	18%
Complex febrile seizures (focal)	29%
Neurodevelopmental abnormalities	33%

278: D. Carbamazepine

Explanation: Management of status epilepticus in children

- Secure Airway, Breathing, Circulation (A, B, C)
- Blood for CBC, electrolytes, glucose, calcium and magnesium
- Give 2ml / kg of a 50% Dextrose i.v. bolus if hypoglycemia
- I/V Lorazepam 0.1mg / kg stat (@ 2mg / min.) or Diazepam 0.3mg / kg stat (@ 1mg / min) followed by

Loading Dilantin (Phenytoin) 20 mg / kg stat
↓ No response even after 15 minutes
Loading Phenobarbitone 15mg / kg stat
↓ No response even after 15 minutes
Midazolam drip 0.2mg / kg stat then 0.2mg / kg / hr. Increase by 0.1mg / kg max. upto 2mg/kg/hr
Alternatively Diazepam i.v. infusion
↓ No respose
Pentobarbitone
Loading 3-5mg / kg followed by 2-3mg / kg / hr [Ventilation, BP, Electrolyte]
↓ Burst suppression EEG x 48 hrs ® stop Pentobarbitone
In refractory status epilepticus, consider GA (Halothane/Thiopentone sodium)

279: A. Valproate

Explanation: **Landau Kleffner Syndrome (LKS)**

- M > F
- Mean age of onset : 5.5 yrs
- Seizure disorder
- Loss of language skills with normal hearing
- Behavioural problem
- EEG : High amplitude spike & wave discharges, bitemporal (non - REM sleep)
- DOC : Na Valproate

280: B. Aqueductal stenosis

Explanation: Causes of hydrocephalus

- Obstructive hydrocephalus: Congenital aqueductal stenosis
- Non-obstructive hydrocephalus: Subarachnoid haemorrhage in western countries, TB meningitis in India

281: C. 10-14 days

Explanation:Therapy for uncomplicated penicillin-sensitive S. pneumoniae meningitis should be for 10-14 days with a third-generation cephalosporin or intravenous penicillin (400,000 units/kg/24 hr, given every 4-6 hr).

282: C. Juvenile myoclonic epilepsy

Explanation: Clues for diagnosis:

- Female adolescent girl
- Positive family history
- Dropping of objects in morning (Early morning myoclonus)
- EEG findings

All this suggests Juvenile Myoclonic Epilepsy, characterised by myoclonus in awake state, with onset in adolescent period and EEG showing 4 to 6 Hz polyspike and slow wave pattern.

283: B. Oral diazepam 6th hourly

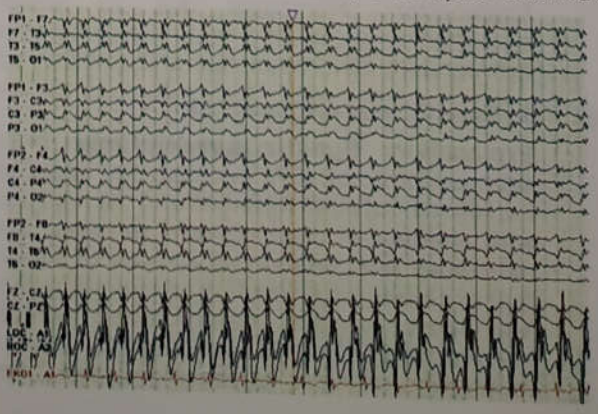
Explanation: Oral BZDs are drugs of choice for prophylaxis of febrile seizures. Remember that paracetamol is not used in prophylaxis.

284: B. Phenytoin

Explanation: First line drugs in absence seizures: Ethosuximide, Valproate
Second line drugs in absence seizures: Clonazepam, Topiramate, Lamotrigine

285: A. 3 Hz spike and wave

Explanation:
EEG: Typical absence seizure, 3 Hz spike and wave



286: C. Post-ictal confusion

Explanation: Absence seizures

- Typical absence seizures usually start at 5-8 yr of age
- Characterised by vacant staring look lasting few seconds
- Absence seizures do not have a postictal period and are characterized by immediate resumption of what the patient was doing before the seizure.
- Hyperventilation for 3-5 min can precipitate the seizures and the accompanying 3 Hz spike–and–slow-wave discharges on EEG

287: A. ACTH

Explanation:

- DOC in infantile spasm/west syndrome: ACTH
- DOC in infantile spasm in tuberous sclerosis: Vigabatrin

288: B. Vigabatrin

Explanation: Read explanation of Q.37.

289: C. Carbamazepine

Explanation: Unlike adults, first line drug for partial seizures in children: Carbamazepine and Oxcarbazepine.

290: A. Complex partial seizure

Explanation: **Automatisms** are automatic semi-purposeful movements of the mouth (oral, alimentary such as chewing) or of the extremities (manual, such as manipulating the sheets; leg automatisms such as shuffling, walking). Automatisms are commonly seen in complex partial seizures (CPS)

291: A. Listeria

Explanation: Listeria produces meningitis in neonates, not in a 12 year old.

292: D. Aseptic meningitis

Explanation: Raised cells, raised protein in CSF but normal CSF glucose is seen in aseptic meningitis (viral meningitis). Also remember the following

	Pressure (40-80mm h2o)	Leuckocytes (2-4mm3)	Protein (20-4-mg/dl)	Glucose (40-80mg/dl)
Bacterial meningitis	Increased	100-50000 PMN	100-500 (high)	Low <40
Viral meningitis	N or increased	Less than 1000 Mononudear	50-200 (high)	N or high
Viral meningitis	N or increased	Less than 1000 PMN early, then mononuclear	50-200 (high)	N or high
T.B meningitis	Increased	10-500 Lymphocytes	100-500 (high)	Low <40
Brain absess	Increased	10-200 lymphocytes	100-500	Normal

293: C. 4-6 weeks

Explanation: In Paediatric brain abscess, the combination of vancomycin, a third-generation cephalosporin, and metronidazole is commonly used intravenously. Abscesses secondary to an infected ventriculo-peritoneal shunt may be initially treated with vancomycin and ceftazidime. The duration of antibiotic therapy depends on the organism and response to treatment but is usually 4-6 wk. Ref. Nelson 20th Ed

294: A. Pneumococcus

Explanation: Pneumococcus cause maximum mortality in meningitis, as per Nelson 20th Ed.

295: B. Streptococcus pneumoniae

Explanation: Meningitis due to Streptococcus pneumoniae is common in children who have splenic dysfunction, CSF rhinorrhea and cochlear implants.

296: B. Spastic quadriplegic CP

Explanation: **Spastic quadriplegia** is the most severe form of CP because of marked motor impairment of all extremities and the high association with intellectual disability and seizures. Swallowing difficulties are common as a result of supra-nuclear bulbar palsies, often leading to aspiration pneumonia. The most common lesions seen on pathologic examination or on MRI scanning are severe PVL and multicystic cortical encephalomalacia. Neurologic examination shows increased tone and spasticity in all extremities, decreased spontaneous movements, brisk reflexes, and plantar extensor responses.

297: B. BERA

Explanation: Children who develop meningitis due to Hemophilus influenzae are prone to develop sensorineural deafness and prior to discharge or on first follow-up, doing an audiometric test is recommended, like BERA (Brainstem evoked response audiometry).

298: C. Severity of myotonia is determined by degree of weakness in muscles

Explanation: Key points regarding myotonic dystrophy (Ref. Nelson 20th Ed)

- Autosomal dominant disorder
- The facial appearance is characteristic, consisting of an inverted V-shaped upper lip, thin cheeks, and scalloped, concave temporalis muscles
- The distal distribution of muscle wasting in myotonic dystrophy is an *exception* to the general rule of myopathies having proximal and neuropathies having distal distribution patterns. The muscular atrophy and weakness in myotonic dystrophy are slowly progressive throughout childhood and adolescence and continue into adulthood
- **Myotonia,** a characteristic feature shared

by few other myopathies, does not occur in infancy and is usually not clinically or even electromyographically evident until about age 5 yr. Exceptional patients develop it as early as age 3 yr. Myotonia is a very slow relaxation of muscle after contraction, regardless of whether that contraction was voluntary or was induced by a stretch reflex or electrical stimulation. The severity of myotonia does not necessarily parallel the degree of weakness, and the weakest muscles often have only minimal myotonia.

- **Cardiac involvement** is usually manifested as heart block in the Purkinje conduction system and arrhythmias (and sudden death)
- **Endocrine abnormalities** involve many glands and appear at any time during the course of the disease. Hypothyroidism is common.
- **Immunologic deficiencies** are common in myotonic dystrophy. The plasma immunoglobulin G level is often low.

299: A. Duchenne muscular dystrophy

Explanation: Calf hypertrophy may be "pseudohypertrophy", positive gower signindicates lower limb girdle weakness and raised CPK levels in a male with weakness in lower limbs is indicative of Duchenne muscular hypertrophy. Nelson 20th Ed says "The serum CK level is consistently greatly elevated in DMD, even in pre-symptomatic stages, including at birth. The usual serum concentration is 15,000-35,000 IU/L (normal <160 IU/L)."

300: D. Duchenne's muscular dystrophy

Explanation: A potential treatment under investigation for duchenne's muscular dystrophy is intravenous or subcutaneous injection of antisense oligonucleotide drugs that induce exon skipping during mRNA splicing in patients with susceptible mutations (~15% of patients) to restore the open reading frame in the DMD gene. Drisapersen and eteplirsen are exon 51 skipping antisense oligonucleotides that bind RNA and skip (bridge) over the defective exon, thus producing a shorter but potentially functional dystrophin protein.

301. Not a B-cell defect?
A. CVID
B. Chronic mucocutaneous candidiasis
C. Hyper IgM syndrome
D. X-linked agammaglobulinemia

302. Most common primary immunodeficiency is?
A. Common variable immunodeficiency
B. Isolated IgA immunodeficiency
C. Wiskott Aldrich syndrome
D. AIDS

303. A child with delayed separation of umbilical cord, leucocytosis and recurrent infections will have?
A. Fulminant neonatal sepsis
B. Leucocyte adhesion defect
C. Histiocytosis-X
D. Myeloperoxidase deficiency

304. Nitroblue tetrazolium test is done for?
A. Phagocytes B. Complement
C. T cells D. B cells

305. Inheritance of Chediak-Higashi syndrome is?
A. X-linked dominant B. X-linked recessive
C. Autosomal dominant D. Autosomal recessive

306. Not seen in DiGeorge syndrome?
A. Decreased T-cells
B. Increased B-cells
C. Absent lymphoid follicles
D. Absent thymus

307. Which variety of SCID shows greatest degree of lymphopenia?
A. Artemis deficiency B. ADA deficiency
C. CD45 deficiency D. X-linked SCID

308. FOXP3 gene mutation is not associated with?
A. Immune dysregulation B. Seizures
C. Polyendocrinopathy D. Enteropathy

309. A 3year old child presents with recurrent pneumonia, eczema and thrombocytopenia. Which protein synthesis will be abnormal in this child?
A. Hamartin B. Wasp
C. Adenosine deaminase D. HLA-1a

310. In MPO deficiency, the increased susceptibility to infections is due to?
A. Defective prostaglandin production
B. Defective rolling of neutrophils
C. Inability to produce hydroxyl halide radicals
D. Inability to produce hydrogen peroxide

311. Hereditary angioneurotic edema is due to deficiency of?
A. C1 inhibitor B. S protein
C. C3b inactivator D. C2 kinins

312. Vitamin C may be useful in the management of which immunodeficiency?
A. MPO deficiency
B. Chediak Higashi syndrome
C. Joubert syndrome
D. DiGeorge syndrome

313. Which among the following B-cell disorder is mostly asymptomatic?
A. CVID
B. Selective IgA deficiency
C. Selective IGM deficiency
D. Pan-agammaglobulinemia

314. Gastrointestinal involvement similar to Crohn's disease is seen in which among the following immunodeficiency disorders?
A. MPO deficiency B. Hyper IGM syndrome
C. CVID D. CGD

315. Increased susceptibility to EBV infection is seen in?
A. Duncan disease B. CVID
C. MPO deficiency D. Kostmann syndrome

316. Hyper IgM syndrome has defect in?
A. Antibody production
B. Antibody class switch recombination
C. Antibody catabolism
D. Bone marrow differentiation

317. Not seen in ataxia telangiectasia?
A. Telangiectasia since birth
B. Ataxia in childhood
C. Low IgE levels
D. Absent IgA

318. Patients with properdin deficiency in complement system are predisposed to infections by?
A. EBV B. Neisseria
C. Staphylococcus aureus D. Giardia

319. A neonate is found to have bloody stools with ear discharge and rash. What should you suspect?
A. DiGeorge syndrome
B. Wiskott Aldrich syndrome
C. Leukocyte adhesion defects
D. Fungal sepsis

320. Most effective screening test for suspected complement defects is?
A. Serum C4 levels B. Serum C3 levels
C. CH50 levels D. None of the above

301. b	302. b	303. b	304. a	305. d	306. c	307. b	308. b	309. b	310. c
311. a	312. b	313. b	314. d	315. a	316. b	317. a	318. b	319. b	320. c

301: B. Chronic mucocutaneous candidiasis

Explanation: Types of primary immunodeficiency diseases

B cell defects	T cell defects	Combined B and T cell defects	Phagocytosis defects
• X-linked agammaglobulinemia • Common variable • Immune deficiency (CVID) • Selective IgA deficiency • Selective IgM/IgG deficiency • Hyper IgM syndrome • Transcobalamin-II deficiency	• Thymic hypoplasia (in DiGeorge syndrome) • Chronic mucocutaneous candidiasis • PNP deficiency	• Severe combined immunodeficiency (SCID) • a. Adenosine deaminase deficiency • b. Reticular dysgenesis of de Vaal • c. Swiss type agamma-globulinemia • • Ataxia tengiectasia • Wiskott Aldrich syndrome • Nezelof Syndrome	• Chronic granulomatous disease • Chediak-higashi syndrome • MPO deficiency • Job syndrome • Hyper IgE syndrome • Lazy leucocyte syndrome • Schwachman's disease • Job's syndrome • Tuftsin deficiency

302: B. Isolated IgA immunodeficiency

Explanation: Isolated IgA immunodeficiency is the MC primary immunodeficiency, with low levels of serum and secretory IgA.

303: B. Leucocyte adhesion defect

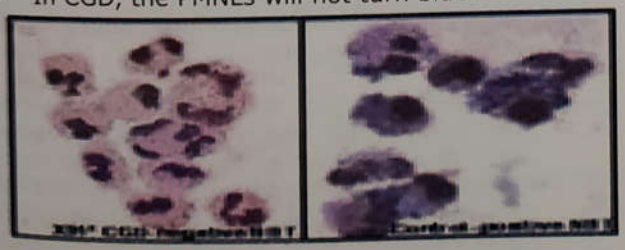
Explanation: According to Nelson 20th Ed, childrenwith severe forms of LAD present in infancy with recurrent,indolent bacterial infections of the skin, mouth, respiratory tract, lowerintestinal tract, and genital mucosa. Significant neutrophilic leukocytosis,often >25,0000/mm3, is a prominent feature. They may have ahistory of delayed separation of the umbilical cord, usually with associatedinfection of the cord stump.

304: A. Phagocytes

Explanation:

Nitro Blue-Tetrazolium Test (NBT)

- Semiquantitative assay
- Tests the phagocytio function or polymorphonclear leukocytes (PMNL)
- Cells reduce a coloriess NBT into a blue-black deposit within the cell
- In CGD, the PMNLs will not turn blue.



305: D. Autosomal recessive

Explanation: Chédiak-Higashi syndrome (CHS) is a rare autosomal recessive disordercharacterized by increased susceptibility to infection caused bydefective degranulation of neutrophils, a mild bleeding diathesis, partial oculocutaneous albinism, progressive peripheral neuropathy,and a tendency to develop a life-threatening form of hemophagocyticlymphohistiocytosis.

306: C. Absent lymphoid follicles

Explanation: Findings in DiGeorge syndrome

- Absolute lymphocyte counts are usually only moderately low for age.
- The CD3 T-cell counts are variably decreased in number, correspondingto the degree of thymic hypoplasia, resulting in an increased percentageof B cells.
- Lymphocyte responses to mitogen stimulation areabsent, reduced, or normal, depending on the degree of thymic deficiency.
- Thymic tissue is absent or hypoplastic. However, when found, contains Hassall corpuscles, anormal density of thymocytes, and corticomedullary distinction.
- Lymphoidfollicles are usually present, but lymph node paracortical areasand thymus-dependent regions of the spleen show variable degrees ofdepletion.

307: B. ADA deficiency

Explanation: Patientswith adenosine deaminase (ADA) deficiency have the lowest absolutelymphocyte counts, usually <500/mm3, but infants with all moleculartypes of SCID are lymphopenic because they lack T cells (normallyaccounting for 70% of circulating lymphocytes).

308: B. Seizures

Explanation: Immune-dysregulation, polyendo-crinopathy, enteropathy, X-linked(IPEX) syndrome is caused by a mutation in the FOXP3 gene, whichencodes a forkhead-winged helix transcription factor (scurfin)involved in the function and development of CD4+CD25+ regulatoryT cells. The absence of regulatory cells may predispose to abnormalactivation of effector T cells.

309: B. Wasp

Explanation: Wiskott-Aldrich syndrome, an X-linked recessive syndrome, is characterizedby atopic dermatitis, thrombocytopenic purpura withnormal-appearing megakaryocytes but small defective

platelets, andundue susceptibility to infectionThe abnormal gene, on the proximal arm of the X chromosome atXp11.22-11.23 near the centromere, encodes a 501 amino acid prolinerichcytoplasmic protein known as the Wiskott-Aldrich syndrome protein (WASP).

310: C. Inability to produce hydroxyl halide radicals

Explanation: Self-explanatory

311: A. C1 inhibitor

Explanation: HAE is a debilitating disease characterized by sudden attacks of brawny, non-pitting, and often painful edema. It is caused either by absolute deficiency or improper function of C1 esterase inhibitor in plasma.

312: B. Chediak Higashi syndrome

Explanation: According to Nelson 20th Ed, high-dose ascorbic acid (200 mg/day for infants, 2,000 mg/day foradults) may improve the clinical status of some children in the stablephase of Chediak-Higashi syndrome. Remember that HSCT is the therapy of choice in these children.

313: B. Selective IgA deficiency

Explanation:

Phenotype	Main Clinical Features
Pan-agammaglobulinemia (absence of lgM, lgG, and lgA)	Bacterial infections (in the respiratory tract) and enterovirus infections
Variable pan-hypogamma-globulinemia (CVID)	Bacterial infections (in the respiratory tract and gut), autoimmunity, cancer and increased risk of granuloma
CSR deficiencies (ebsence or decrease in levels of lgG and lgA)	Bacterial and opportunistic infections
Selective lgA deficiency	Most often asymptomatic
Selective lgM deficiency	Frequent infections with encapsulated bacteria

314: D. CGD

Explanation: Granuloma formation and inflammatory processesare a hallmark of CGD and may be the presenting symptomsthat prompt testing for CGD if they cause pyloric outlet obstruction,bladder outlet or ureter obstruction, or rectal fistulas and granulomatouscolitis simulating Crohn disease. More than 80% of CGD patientshave positive serology for Crohn disease.

315: A. Duncan disease

Explanation: X-linked lymphoproliferative disease (XLP disease), also referred to as Duncan disease after the originalkindred in which it was described, is an X-linked recessive traitcharacterized by an inadequate immune response to infection withEpstein-Barr virus (EBV).

316: B. Antibody class switch recombination

Explanation: The hyper-IgM syndrome is genetically heterogeneous and characterized by normal or elevated serum IgM levels associated with lowor absent IgG, IgA, and IgE serum levels, indicating a defect in theclass-switch recombination (CSR) process.

317: A. Telangiectasia since birth

Explanation: In Ataxia telangiectasia, the most prominent clinical features are progressive cerebellar ataxia,oculocutaneous telangiectasias, chronic sinopulmonary disease, ahigh incidence of malignancy, and variable humoral and cellularimmunodeficiency. Ataxia typically becomes evident soon after thesechildren begin to walk and progresses until they are confined to awheelchair, usually by the age of 10-12 yr. The telangiectasias begin todevelop at 3-6 yr of age. The most frequent humoral immunologicabnormality is the selective absence of IgA, which occurs in 50-80% ofthese patients. Hypercatabolism of IgA also occurs. IgE concentrationsare usually low, and the IgM may be of the low-molecular-weightvariety. IgG2 or total IgG levels may be decreased, and specific antibodytiters may be decreased or normal.

318: B. Neisseria

Explanation: Persons with properdin deficiency have a striking predisposition toN. meningitidis meningitis.

319: B. Wiskott Aldrich syndrome

Explanation:

Features	Diagnosis
In newborn and young infants (0-6 MO)	
Hypocalcemia, unusal facies and ears, heart disease	DiGeorge anomaly
Delayed umbilical cord detachment, leukocytosis, recurrent infections	Leukocyte adhesion defect
Presistent thrush, failure to thrive, pneumonia, diarrhea	Severe combined immuno deficiency
Blood stools, draining ears, atopic eczema	Wiskott-Aldrich syndrome
Pneumocystic jiroveci pneumonia, neutropenia, recurrent infections	X-linked hyper-lgM syndrome

320: C. CH50 levels

Explanation: The most effective screening test for complement defects is a CH50assay, which is a bioassay that measures the intactness of the entirecomplement pathway and yields abnormal results if complementhas been consumed from the specimen for any reason. Genetic deficienciesin the complement system are usually characterized byextremely low CH50 values.

321. Which of the following is a preventable cause of mental retardation?

A. Hypothyroidism
B. Down's syndrome
C. Cerebral palsy
D. All of the above

322. The most common cause of ambiguous genitalia in a newborn is:

A. 21 hydroxylase deficiency
B. 11 beta hydroxylase deficiency
C. 17 alpha hydroxylase deficiency
D. 3 beta hydroxysteroid dehydrogenase deficiency

323. A male child with 3 beta-hydroxysteroid dehydrogenase deficiency cannot have which among the following?

A. Hypoglycaemia
B. Normal genitalia
C. Increased DHEA
D. Decreased androstenedione

324. The first ophthalmic evaluation in children with type 2 DM is performed at?

A. At the time of diagnosis
B. 2 yr after diagnosis is made
C. 5 yr after diagnosis is made
D. 10 yr after diagnosis is made

325. Most common cause of Addison disease in children is?

A. Idiopathic
B. Autoimmune destruction
C. Infectious causes
D. Tubercular

326. A 10 month old female baby, previously normal, suddenly becomes distressed in crib. External genitalia is normal, except some hyperpigmentation. Blood glucose is 30 mg%. Most likely diagnosis?

A. CAH due to 21 hydroxylase deficiency
B. Hyperinsulinism
C. Familial glucocorticoid deficiency
D. Cushing syndrome

327. A 5-year old boy with precocious puberty and well developed pubic hair with normal genital and BP 120/90, the following would be seen to be deficient?

A. Deoxycorticosterone
B. 11 beta hydroxylase
C. 17 alpha hydroxylase
D. Aldosterone

328. Which one of the following drugs is used in the fetal treatment of CAH?

A. Hydrocortisone
B. Prednisolone
C. Fludrocortisone
D. Dexamethasone

329. Drug of choice for CAH?

A. Hydrocortisone
B. Prednisolone
C. Fludrocortisone
D. Dexamethasone

330. Bone age advanced of chronological age is seen in?

A. GH deficiency
B. Precocious puberty
C. Hypothyroidism
D. Constitutional delay in growth

331. Not seen in Sotos syndrome?

A. Macrocephaly
B. Hypertelorism
C. Mongoloid slant of eyes
D. Ataxic gait

332. Which endocrine disorders is associated with epiphyseal dysgenesis?

A. Hypothyroidism
B. Cushing's syndrome
C. Addison's disease
D. Hypoparathyroidism

333: A 5 year old female child presents with virilization and hypertension, with decreased renin levels and with hypernatremia and hypokalemia, diagnosis is?

A. 11 beta hydroxylase deficiency
B. 3 beta hydroxy steroid dehydrogenase def.
C. 21 alpha hydroxylase
D. Conn syndrome

334: Inheritance in congenital adrenal hyperplasia is?

A. Sporadic
B. Mitochondrial
C. Autosomal dominant
D. Autosomal recessive

335. Most common cause of endogenous cushing syndrome in a 10 year old child is?

A. Glucocorticoid ingestion
B. Pituitary adenoma
C. McCune Albright syndrome
D. Tumours

336. In Paediatric age, HbA1C cut off (in g/dl) to define diabetes mellitus is?

A. 6.0
B. 6.5
C. 7.0
D. 7.5

337. Which congenital infection has maximum risk of type 1 diabetes mellitus?

A. Congenital CMV
B. Congenital toxoplasmosis
C. Congenital rubella
D. Congenital syphilis

338. Dawn phenomenon in T1 DM is mediated by?

A. Overnight GH secretion
B. Decreased cortisol activity
C. Decreased insulin clearance
D. Increased cortisol activity

339. Glucokinase deficiency is associated with which variety of MODY?

A. MODY1
B. MODY2
C. MODY3
D. MODY4

340. All of the following are seen in Donohue syndrome, except?

A. Fasting hypoglycaemia
B. Postprandial hyperglycemia
C. Normal life expectancy
D. Insulin resistance

321. a	322. a	323. b	324. a	325. b	326. c	327. b	328. d	329. a	330. b
331. c	332. a	333. a	334. d	335. b	336. b	337. c	338. a	339. b	340. c

321: A. Hypothyroidism

Explanation: Hypothyroidism is the most common preventable cause of mental retardation

322: A. 21 hydroxylase deficiency

Explanation: The most common cause of ambiguous genitalia in a neonate is Congenital adrenal hyperplasia (CAH). The most common cause of CAH is 21 hydroxylase deficiency, followed by 11 beta hydroxylase deficiency.

323: B. Normal genitalia

Congenital adrenal hyperplasia due to 3-beta hydroxy steroid dehydrogenase deficiency always produces ambiguous genitalia in both males and females.

Quick review:

Features of 3-beta hydroxy steroid dehydrogenase deficiency and hormonal picture (Nelson 20th Ed)

Glucocorticoid deficiency	↓ Cortisol, ↑ ACTH ↑ ↑ Baseline and ACTH stimulated ▲5 steroids (pregnenolone, DHEA)
Mineralocorticoid deficiency (salt-wasting crisis)	Hyponatremia, hyperkalemia ↑ Plasma renin
Ambiguous genitalia in females and males	↑ DHEA, ↓ androstenedione, testosterone, and estradiol
Precocious adrenarche, disordered puberty	↑ DHEA, ↓ androstenedione, testosterone, and estradiol

324: A. At the time of diagnosis

Explanation: As per Nelson 20th Ed, all childhood diabetic patients need to have an initial dilated and comprehensive examination by an ophthalmologist shortly after the diagnosis of diabetes is made in patients with T2DM, and within 3-5 yr after the onset of T1DM (but not before age 10 yr). Any patients with visual symptoms or abnormalities should be referred for ophthalmologic evaluation. Subsequent evaluations for both T1DM and T2DM patients should be repeated annually by an ophthalmologist who is experienced in diagnosing the presence of diabetic retinopathy and is knowledgeable about its management.

325: B. Autoimmune destruction

Explanation: One-liner

326: C. Familial glucocorticoid deficiency

Explanation: Familial glucocorticoid deficiency is a form of chronic adrenal insufficiency characterized by isolated deficiency of glucocorticoids, elevated levels of ACTH, and generally normal aldosterone production, although salt-losing manifestations as are present in most other forms of adrenal insufficiency occasionally occur. Patients mainly have hypoglycemia, seizures, and increased pigmentation during the 1st decade of life. The disorder affects both sexes equally and is inherited in an autosomal recessive manner.

327: B. 11 beta hydroxylase

Explanation: In 11 beta hydroxylase deficiency, approximately65% of patients become **hypertensive** due to elevated deoxycorticosterone, which has mineralocorticoid action. Ambiguous genitalia occurs in females and males have normal genitalia with precocious puberty.

328: D. Dexamethasone

Explanation: Motherswith pregnancies at risk are given dexamethasone, a steroid that readilycrosses the placenta, in an amount of 20 μg/kg pre-pregnancy maternalweight daily in 2 or 3 divided doses. This suppresses secretion of steroidsby the fetal adrenal, including secretion of adrenal androgens. Ifstarted by 6 wk of gestation, it ameliorates virilization.

329: A. Hydrocortisone

Explanation: DOC for postnatal therapy of CAH is Hydrocortisone

330: B. Precocious puberty

Explanation: **Osseous maturation** is variably advanced, often by more than 2-3SD in precocious puberty.

331: C. Mongoloid slant of eyes

Explanation: Clinically,sotos syndrome is characterized by a large (macrocephaly), dolichocephalic head, prominent forehead and jaw, hypertelorism,antimongoloid slant of the palpebral fissures, high-arched palate, andlarge hands and feet with thickened subcutaneous tissue. Clumsinessand awkward gait are also noted. Seizures and some intellectual dysfunction may also occur.

332: A. Hypothyroidism

Explanation: One-liner

333: A. 11 beta hydroxylase deficiency

Explanation: Salt retaining crisis with female virilization is seen in 11 beta hydroxylase deficiency.

334: D. Autosomal recessive

Explanation: All varieties of CAH are autosomal recessive.

335: B. Pituitary adenoma

Explanation: The most common etiology ofendogenous Cushing syndrome in children older than 7 yr of age is**Cushing disease,** in which excessive ACTH secreted by a pituitaryadenoma causes bilateral adrenal hyperplasia.

336: B. 6.5

Explanation: As per the latest update in 2018 by the American Diabetes Association (ADA), the diagnostic criteria for diabetes mellitus includes:

i. Fasting plasma glucose (FPG) ≥ 126 mg/dL (7.0 mmol/L). Fasting is defined As no caloric intake for at least 8 hours

OR

ii. Two-hours plasma glucose ≥ 200 mg/dL (11.1 mmol/L) during an oral glucose tolerance test (OGTT). The test should be performed as described by the WHO, using a glucose load containing the equivalent of 75 g anhydrous glucose dissolved in water

OR

iii. Hb A1C ≥ 6.5% (48 mmol/mol). The test should be performed in a laboratory using a method that is NGSP-certified and standardized to the DCCT assay

OR

iv. In a patient with classic symptoms of hyperglycemia or hyperglycemic crisis, a random plasma glucose ≥ 200 mg/dL (11.1 mmol/L).

337: C. Congenital rubella

Explanation: According to Nelson 20th Ed, the clearest evidence of a role for viral infection in human T1DM isseen in congenital rubella syndrome. Prenatal infection with rubella isassociated with β-cell autoimmunity in up to 70%, with developmentof T1DM in up to 40% of infected children. The time lag betweeninfection and development of diabetes may be as high as 20 yr.

338: A. Overnight GH secretion

Explanation: There are several reasons that blood glucose levels increase in the earlymorning hours before breakfast. The most common is a simple declinein insulin levels. This usually results in routinely elevated morningglucose. The **dawn phenomenon** is thought to be mainly caused byovernight growth hormone secretion and increased insulin clearance. It is a normal physiologic process seen in most adolescents withoutdiabetes, who compensate with more insulin output. A child withT1DM cannot compensate. The dawn phenomenon is usually recurrentand modestly elevates most morning glucose levels.

339: B. MODY2

Explanation: Types of MODY

	Gene Mutated	Function
Mody1	HNF4α	Transcription factor
Mody2	Glucokinase (GCK)	Enzyme, glucose sensor
Mody3	HNF1α	Transcription factor
Mody4	IPF-1	Necessary for pancreatic development
Mody5	HNF1β	Transcription factor
Mody6	NEUROD1	Differentiation factor in the development of pancreatic islets
Mody7	KFL11	Zinc finger transcription factor
Mody8	CEL	Bile salt-dependent lipase
Mody9	PAX4	Transcription factor
Mody10	INS	Insulin gene
Mody11	BLK	B-lymphocyte tyrosine kinase

340: C. Normal life expectancy

Explanation: Donohue Syndrome

- This is a syndrome characterized by intrauterine growth restriction,fasting hypoglycemia, and postprandial hyperglycemia in associationwith profound resistance to insulin; severe hyperinsulinemia is seencompared to age-matched infants during an oral glucose tolerance test.
- Various defects of the insulin receptor have been described, therebyattesting to the important role of insulin and its receptor in fetal growthand possibly in morphogenesis.
- Most of these patients die in the 1st yrof life.

341. Biomarker not involved in acute kidney injury is?

A. NGAL
B. KIM-1
C. Micro RNA 122
D. Cystatin-C

342. An 8-year-old child suffering from recurrent attacks of polyuria since childhood presents to OPD. On examination, he has short stature, normal vitals and normal BP. Serum creatinine 6 mg%, HCO3 is 16 mEq, Na^+ is 134, K^+ is 4.2 and ultrasonography shows bilateral small kidneys. What is the diagnosis?

A. Juvenile nephronophthisis
B. Reflux nephropathy
C. Polycystic kidney disease
D. Medullary cystic kidney disease

343. A 7-year-old child has first episode of nephrotic syndrome one year back, which was inappropriately treated with low dose corticosteroids for 4 weeks. Since then, he had 4 more documented relapses and every time, the child responded to oral prednisolone. What is the best management for this child?

A. Pulse methylprednisolone
B. Start Levamisole
C. Start Cyclophosphamide
D. Start Cyclosporine

344. A 2-month-old girl has failure to thrive, polyuria and medullary nephrocalcinosis affecting both the kidneys. Investigations showed blood pH of 7.48, bicarbonate of 25 mEq/l, potassium of 2 meq/l, sodium of 126 meq/l and chloride of 88 meq/l. The most likely diagnosis is?

A. Distal renal tubular acidosis
B. Primary hyperaldosteronism
C. Barter syndrome
D. Pseudohypoaldosteronism

345. A 10 years old girl presented with hypokalemia, mild hypercalciuria, hypertension and metabolic alkalosis. Which among the following is the likely diagnosis?

A. Bartter syndrome
B. Gitelman syndrome
C. Liddle syndrome
D. Alport syndrome

346. A known case of childhood nephrotic syndrome presents with fever and sudden onset severe pain abdomen with a palpable mass in lower left abdomen. Likely diagnosis is?

A. Spontaneous bacterial peritonitis
B. Renal vein thrombosis
C. Chronic pyelonephritis
D. Acute renal failure

347. A 5-year-old child was diagnosed to have steroid-resistant nephrotic syndrome, with no response to oral prednisolone at 2 mg/kg/day despite 28 days of daily administration. What is the next treatment?

A. IV methylprednisolone
B. IV cyclophosphamide
C. Oral Levamisole
D. Oral cyclosporine

348. A child successfully treated for 1st episode of nephrotic syndrome is advised pneumococcal vaccination. What is the earliest time he can be given that?

A. 2 weeks after last steroid dose
B. 4 weeks after last steroid dose
C. 6 weeks after last steroid dose
D. 8 weeks after last steroid dose

349. Drug of choice for Frequently relapsing nephrotic syndrome?

A. Methylprednisolone
B. Cyclophosphamide
C. Cyclosporine
D. Rituximab

350. Most common cause of renal vein thrombosis in children?

A. Minimal change disease
B. PDA
C. Dehydration
D. FSGS

351. All of the following genes are involved in congenital nephrotic syndrome, except?

A. WT1
B. WT2
C. LMX1B
D. SMARCAL1

352. Most common cause of UTI in children?

A. Klebsiella
B. E coli
C. Proteus
D. Pseudomonas

353. Which among the following is the most common cause of gross hematuria in children?

A. PSGN
B. Renal stones
C. UTI
D. IgA nephropathy

354. A child aged 13 months is successfully treated for UTI using antibiotics and comes on first follow-up visit. What is the next investigations to perform?

A. USG-KUB
B. USG-KUB + DMSA
C. USG-KUB + DMSA + MCU
D. DMSA + MCU

355. A child with recurrent UTI is most likely to show

A. PUV
B. VUR
C. Neurogenic bladder
D. Renal and ureteric calculi

356. In HUS associated with shigella in children, the false statement is:

A. Leucocytosis
B. Neurological abnormalities
C. Hepatic failure
D. Thrombotic angiopathy

341. c	342. a	343. c	344. c	345. c	346. b	347. d	348. b	349. b	350. c
351. b	352. b	353. c	354. b	355. b	356. c				

357. **True regarding minimal change disease?**
A. More common in girls than boys
B. Non-selective proteinuria
C. Severe hypoalbuminemia
D. Cyclosporine is effective first-line therapy

358. **A child aged 6 yr is asymptomatic but incidentally done urine culture showed 1 million colony count/mm3 of E. coli. What is the next step in management?**
A. Repeat urine culture
B. Nitrite test
C. Nothing needed
D. Start oral antibiotics

359. **As per RIFLE staging system criteria, the minimum time duration to define end stage renal disease is?**
A. 4 weeks
B. 8 weeks
C. 3 months
D. 6 months

360. **The most common age for developing acute post-streptococcal glomerulonephritis is?**
A. Below 2 yrs
B. 2-6 yr
C. 6-12 yr
D. >12 yr

361. **A 5 year old child developed periorbital puffiness following an acute febrile illness. His spot urine protein sample showed protein: creatinine ratio of 3.5:1. There was no hypertension or hematuria. The child was started on oral prednisolone @ 2mg/kg/day as single morning dose. Despite 2 weeks of therapy, the child continues to have periorbital edema and persistently elevated protein: creatinine ratio. What next needs to be done?**
A. Continue prednisolone in same dose for 2 weeks more
B. Increase dose of prednisolone to double
C. Start Levamisole as steroid sparing agent
D. Consider renal biopsy

362. **A child develops nephrotic syndrome, which is poorly responsive to prednisolone. He has past history of VUR, Renal biopsy in such a child is likely to show which pathology?**
A. Effacement of foot process of podocytes
B. Focal segmental glomerulosclerosis
C. Subepithelial deposits
D. Mesangial IgA deposits

363. **A child with nephrotic syndrome has been advised to undergo pneumococcal vaccination. When can you start the vaccination?**
A. Immediately after remission is attained
B. Along with daily steroid
C. 4 weeks after last dose of steroid
D. 8 weeks after last dose of steroid

364. **Renal potassium wasting associated with hypertension is seen in all except?**
A. Conn syndrome
B. Cushing syndrome
C. Bartter syndrome
D. Liddle syndrome

365. **The treatment of choice for primary grade V VUR involving both kidneys in a 6-month old boy is?**
A. Antibiotic prophylaxis
B. Ureteric reimplantation
C. Cystoscopy followed by sub-ureteric injection of Teflon
D. Bilateral ureterostomies

366. **A 9-year old boy has steroid dependent NS for the last 5 years. The patient is markedly cushingoid with blood pressure of 120/86mm Hg and has small subcapsular cataracts. The most appropriate therapy of choice?**
A. Long-term prednisolone with enalapril
B. Cyclophosphamide
C. IVIG
D. IV pulse corticosteroids

367. **Most common cause of acute kidney injury in neonates is?**
A. Hypovolemia
B. HUS
C. Sepsis
D. Urinary obstruction

368. **Which among the following is an indication of Renal biopsy in a patient of nephrotic syndrome?**
A. Massive proteinuria
B. Age 2-8 years
C. Low serum complement levels
D. 2nd attack of nephrotic syndrome in 1 year

369. **Most common cause of urinary obstruction a male infant is?**
A. Anterior urethral valves
B. Posterior urethral valves
C. Stone
D. stricture

370. **Which one of the following is the most common cause of abdominal mass in neonates?**
A. Neuroblastoma
B. Wilm'stumor
C. Distended bladder
D. Multicystic dysplastic kidney

357. c	358. c	359. c	360. c	361. a	362. b	363. c	364. c	365. a	366. b
367. c	368. c	369. b	370. d						

341: C. Micro RNA 122

Explanation: The typical marker used to define AKI is Serum creatinine. However, since it is both insensitive and delayed, newer markers to define AKI have come up. These include:

- Plasma cystatin C
- Plasma NGAL
- Urinary IL-18
- Urinary NGAL
- Urinary KIM-1

342: A. Juvenile nephronophthisis

Explanation: Child with polyuria, normal BP, metabolic acidosis (low HCO3) and bilateral small kidneys favours nephronophthisis.

343: C. Start Cyclophosphamide

Explanation: This is a child with frequently relapsing nephrotic syndrome (FRNS). **Cyclophosphamide** prolongs the duration of remission and reduces the number of relapses in children with **frequently relapsing** and **steroid-dependent** nephrotic syndrome. The potential side effects of the drug (neutropenia, disseminated varicella, hemorrhagic cystitis, alopecia, sterility, increased risk of future malignancy) should be carefully reviewed with the family before initiating treatment. Cyclophosphamide (2 mg/kg) is given as a single oral dose for a total duration of 8-12 wk. Alternate-day prednisone therapy is often continued during the course of cyclophosphamide administration.

During cyclophosphamide therapy, the white blood cell count must be monitored weekly and the drug should be withheld if the count falls below 5,000/ mm3. The cumulative threshold dose above which oligospermia or azoospermia occurs in boys is >250 mg/kg.

344: C. Barter syndrome

Explanation:

- Distal RTA has acidosis, not alkalosis
- Primary hyperaldosteronism shows high sodium and hypertension
- Pseudo-hypoaldosteronism shows features of aldosterone deficiency despite high to normal serum aldosterone levels. It shows hyperkalemia.
- The only option left by exclusion is C, which fulfils all the features

345: C. Liddle syndrome

Explanation: Presence of Hypertension, Hypokalemia, Alkalosis and Hypercalciuria is seen only in Liddle syndrome. Remember that Barter syndrome never shows hypertension and Gitelman syndrome shows hypocalciuria.

346: B. Renal vein thrombosis

Explanation: RVT is a known complication of nephrotic syndrome and should be suspected in sudden onset pain abdomen, palpable mass in flanks and/or rising blood urea nitrogen and serum creatinine.

347: D. Oral cyclosporine

Explanation: Cyclosporine is the DOC for SRNS

348: B. 4 weeks after last steroid dose

Explanation: Live vaccines can be used in nephrotic syndrome minimum 4 weeks after the last dose of steroid therapy has been given.

349: B. Cyclophosphamide

Explanation:

- DOC for frequently relapsing and steroid dependent NS: Cyclophosphamide
- DOC for steroid resistant nephrotic syndrome: Cyclosporine/Tacrolimus

350: C. Dehydration

Explanation: Dehydration is the MC cause of renal vein thrombosis in children. In adults, it is mostly due to membranous nephropathy.

351: B. WT2

Explanation:

Genes implicated in congenital nephrotic syndrome:

Gene	Name	Location	Inheritance
Steroid-Resistant Nephrotic Syndrome			
NPHS1	Nephrin	19q13.1	Recessive
NPHS2	Podocin	1q25	Recessive
WT1	Wilms tumor suppressor gene	11p13	Dominant
LMX1B	LIM-homeodomain protein	9q34	Dominant
SMARCAL1	SW / SNF2- related, matrix-associated, actin-dependent regulator of chromatin, subfamily a link-1	2q35	Recessive

352: B. E coli

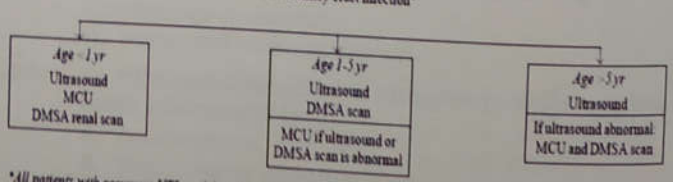
Explanation: MCC of Childhood UTI: E coli, followed by Klebsiella

353: C. UTI

Explanation: According to Nelson 20th Ed, the most common cause of grosshematuria in children is bacterial urinary tract infection.

354: B. USG-KUB + DMSA

Explanation: As per Indian Society of Paediatric Nephrology Guidelines, any child with first episode of UTI needs to be evaluated after completion of antibiotic therapy on first follow-up visit as follows:



*All patients with recurrent UTI need detailed evaluation with ultrasonography, DMSA scan and MCU

355: B. VUR

Explanation: VUR (Vesico-ureteric reflux) is the most common cause of recurrent UTI in children.

356: C. Hepatic failure

Explanation:

- Leucocytosis, raised ESR and CRP may occur in HUS due to inflammation
- The majority of patients with HUS have some central nervous system (CNS) involvement. Most have mild manifestations, with significant irritability, lethargy, or nonspecific encephalopathic features. Severe CNS involvement occurs in ≤20% of cases. Seizures and significant encephalopathy are the most common manifestations in those with severe CNS involvement, resulting from focal ischemia secondary to microvascular CNS thrombosis. Small infarctions in the basal ganglion and cerebral cortex have also been reported, but large strokes and intracranial hemorrhage are rare. Hypertension may produce an encephalopathy and seizures.
- Hepatic failure is not known to occur
- Thrombotic angiopathy involving small vessels in kidneys is a known pathogenetic process

357: C. Severe hypoalbuminemia

Explanation: Minimal change disease is more common in boys than girls, causes selective proteinuria, leading to severe hypoalbuminemia and is thus the MC cause of childhood nephrotic syndrome. Most cases respond well to Oral Prednisolone as the first line therapy.

358: C. Nothing needed

Explanation: This child has asymptomatic bacteriuria. It needs no therapy in any age, except pregnant females

359: C. 3 months

Explanation: Paediatric RIFLE staging

Stage	eCCI%	Urine output
R= Risk for renal dysfunction	eCCI decrease by 25	<0.5 ml/kg/h for 8 h
I= Injury to the kidney	eCCI decrese by 50	<0.5 ml/kg/h for 16h
F= Failure of kidney	eCCI decrease by 74 or <35 ml/min/1.73m^2	<0.3 ml/kg/h for 24h or anuria for 12h
L= Loss of kidney function presistent failure >4 weeks		
E= ESRD persistent failure >3 months		

eCCI: Estimated creatinine clearancel ESRD: End-stage renal disease; RIFLE: Risk injury, loss, end; pRIFL: Paediatric risk, injury, failure, loss, end

360: C. 6-12 yr

Explanation: Poststreptococcal GN is most common in children ages 5-12 yr and uncommon before the age of 3 yr. The typical patient develops an acute nephritic syndrome 1-2 wk after an antecedent streptococcal pharyngitis or 3-6 wk after a streptococcal pyoderma.

361: A. Continue prednisolone in same dose for 2 weeks more

Explanation: This child has nephrotic syndrome with minimal change disease as likely cause (since there is no hypertension or hematuria). Child has been started on optimal dose of corticosteroids and need top be given for 8 weeks before we can label it as steroid resistant or consider biopsy. Steroid sparing agents are not useful in minimal change disease.

362: B. Focal segmental glomerulosclerosis

Explanation: Recurrent UTI & VUR can produce renal scarring in the kidney. If such a patient evolves into nephrotic syndrome, the histological correlate is usually of Focal segmental glomerulosclerosis (FSGS).

363: C. 4 weeks after last dose of steroid

Explanation: Children with nephrotic syndrome additionally need Pneumococcal and varicella vaccination in all cases. Vaccination is done 4 weeks after the last steroid dose.

364: C. Bartter syndrome

Explanation: A child with Bartter syndrome is always normotensive!

Bartter syndrome is characterized by tubular dysfunction, manifesting as Hypokalemia, metabolic alkalosis, secondary hyperaldosteronism, JGA hyperplasia but normal BP.

365: A. Antibiotic prophylaxis

Explanation: In case of VUR, as per changed guidelines, antibiotic prophylaxis is recommended for any grade VUR. Indications of surgery include breakthrough UTI on continuous antibiotic prophylaxis or bladder outlet obstruction.

366: B. Cyclophosphamide

Explanation: For SDNS with features of steroid toxicity, the next drug is Levamisole and best drug is Cyclophosphamide

367: C. Sepsis

Explanation: Most common causes of Acute kidney injury (AKI) in children:

- Neonates: Sepsis, followed by hypovolemia
- Infants: Hypovolemia
- Older children: HUS

368: C. Low serum complement levels

Explanation: Indications of renal biopsy in Nephrotic syndrome

- Suspected secondary cause/NS with significant lesion
- Steroid resistant nephritic syndrome
- Prior to starting Calcineurin inhibitors
- 2 years after starting calcineurin inhibitors
- Low serum complement levels
- Presence of acute renal failure in Nephrotic syndrome
- Congenital NS

369: B. Posterior urethral valves

Explanation: One-liner

370: D. Multicystic dysplastic kidney

Explanation: MCC of abdominal mass in neonates: Multicystic dysplastic kidney

MC malignant cause of abdominal mass in neonates: Neuroblastoma

371. The most common leukocytoclastic vasculitis affecting children is?

A. Takayasu disease
B. Mucocutaneous lymph node syndrome (Kawasaki disease)
C. Henoch-SchoenleinPurpura
D. Polyarteritisnodosa

372. Indication of Prednisolone in Kawasaki disease?

A. Giant coronary aneurysms
B. Age <1 year
C. Massive thrombocytosis with ESR> 50/hr
D. Failure to make patient afebrile despite 2 courses of IVIG

373. What is the recurrence rate of Kawasaki disease?

A. 0-1%
B. 1-3%
C. 3-10%
D. 10-35%

374. Most common type of juvenile idiopathic arthritis is?

A. Polyarticular
B. Enthesitis-related
C. Oligoarticular
D. Systemic-onset JIA

375. Henoch-schonlein purpura commonly involves which age group?

A. 4-8 years
B. 10-15 years
C. 15-20 years
D. Above 20 years

376. A 2 year old child was diagnosed as having Kawasaki disease and was given IVIG in a dose of 2 g/kg. 36 hr later, the child continues to be febrile and has significant conjunctival and mucosal erythema. What next needs to be done?

A. Wait and watch
B. Repeat dose of IVIG
C. Start corticosteroids
D. Start Etanercept

377. Not seen in henochschonlein purpura?

A. Purpura on buttocks
B. Renal involvement
C. Arthritis
D. Thrombocytopenia

378. All of the following are features to diagnose Kawasaki disease, except?

A. Myocarditis
B. High fever lasting atleast 5 days
C. Desquamation and rash
D. Erythematous changes in the oral mucosa

379. Drug of choice in Kawasaki disease?

A. Prednisolone
B. IVIG
C. Rituximab
D. Infliximab

380. Incidence of coronary artery aneurysms in untreated Kawasaki disease?

A. <5%
B. 5-10%
C. 15-25%
D. 40-60%

381. Juvenile idiopathic arthritis of systemic onset variety, all are seen except?

A. High Fever
B. Salmon coloured rash
C. Hepatosplenomegaly
D. Uveitis in most patients

382. Characteristic feature of systemic onset juvenile idiopathic arthritis?

A. Begins after 16 years of age
B. Uveitis is common
C. RA factor is negative
D. NSAIDs are contraindicated

383. A 5 years old child presents with perivascular IgA deposition and neutrophilic collection. There is erythematous rash on the lower limbs and non-blanching purpura. Probable diagnosis is?

A. Henoch-schonlein purpura
B. Wegener's granulomatosis
C. Cryoglobulinemia
D. Kawasaki disease

384. Which of the following is not seen in Henoch schonlein purpura?

A. Purpura
B. Abdominal pain
C. Seizures
D. Arthralgia

385. Which immunoglobulin gets deposited perivascularly in HSP?

A. IgM
B. IgG
C. IgA
D. IgD

386. The most common causative agent in plant thorn synovitis in children is?

A. Trichinella spiralis
B. Pantoeaagglomerans
C. Enterococci
D. Klebsiella

387. Most common type of scleroderma in childhood is?

A. Diffuse systemic
B. Morphea
C. Bullous
D. Pansclerotic

388. In HSP, renal involvement can occur upto what time of diagnosis?

A. Within 1 month
B. Within 2 months
C. Within 4 months
D. Within 6 months

389. Familial form of sarcoidosis shows which type of inheritance?

A. AR
B. AD
C. XLD
D. XLR

390. Not seen in Kawasaki disease?

A. Fever lasting more than 4 weeks
B. Desquamation after 2-3 weeks of disease onset
C. Raised acute phase reactants
D. High risk of cardiac complications

371. c	372. d	373. b	374. c	375. a	376. b	377. d	378. a	379. b	380. c
381. d	382. c	383. a	384. c	385. c	386. b	387. b	388. d	389. b	390. a

371: C. Henoch-SchoenleinPurpura

Explanation: HSP is the most common vasculitis in children and histopathology shows evidence of leukocytoclastic vasculitis.

372: D. Failure to make patient afebrile despite 2 courses of IVIG

Explanation: Kawasaki disease requires Prednisolone only if patient fails to get afebrile despite 2 courses of IVIG.

373: B. 1-3%

Explanation: The vast majority of patients with KD return to normal health, as timely treatment with IVIG reduces the risk of coronary aneurysms to less than 5%. Acute KD recurs in 1-3% of cases, however.

374: C. Oligoarticular

Explanation:

International league of associations for rheumatology subtype	Peak age of onset (yr)	Female: Male Ratio	Percentage of all jia cases	Arthritis pattern	Extraarticular features
Systemic arthritis	1-5	1:1	5-15	Polyarticular, often affecting kness, wrists and ankles; also fingers, neck and hips	Daily fever; evanescent rash; pericarditis; pleuritis
Oligoarthritis	2-4	3:1	40-50 (but ethnic variation)	Knees ++; ankles, fingers +	Uveitis in = 30% of cases
Polyarthritis: RF-negative	2-4 and 10-14	3:1 and 10:1	20-35	Symmetric or asymmetric; small and large joints; cervical spine; temporomandibular joint	Uveitis in = 10%
RF-positive	9-12	9:1	<10	Aggressive symmetric polyarthritis	Rjei,atpod mpdi;es in 10%; low-grade fever
Psoratic arthritis	2-4 and 9-11	2:1	5-10	Asymmetric arthritis of small or medium-sized joints	Uveitis in 10%; psoriasis in 50%
Enthesitis related arthritis	9-12	1:7	5-10	Predominantly lower limb joints affected; sometimes axial skeleton (but less than in adult, ankylosing spondylitis)	Acute anterior uveitis; association with reactive arthritis and inflammatory bowel disease

375: A. 4-8 years

Explanation: Henoch schonlein purpura is most commonly seen in young children between 3 to 10 years of age. It is rare to see HSP below 1 year or above 20 years, although some cases have been reported.According to Nelson 20th Ed, "approximately 90% of HSP cases occur in children, usually between the ages of 3 and 10 yr. HSP is distinctly less common in adults, in whom severe and chronic complications are often encountered."

376: B. Repeat dose of IVIG

Explanation: **The patient has IVIG-resistant KD,** whichoccurs in approximately 15% of patients and is defined by persistent or recrudescent fever 36 hr after completion of the initial IVIG infusion. Patients with IVIG resistance are at increased risk for CAA. Typically, another dose of IVIG at 2 g/kg is administered to patients with IVIG resistance. Corticosteroids in varying doses and via different routes have also been used as secondary or "rescue" therapy when fever persists after the first IVIG. Tumor necrosis factor inhibitors, including infliximab and etanercept, have also been given for the treatment of IVIG-resistant disease in steroid non-responsive cases.

377: D. Thrombocytopenia

Explanation: Non-thrombocytopenic purpura is seen in HSP in dependent areas like buttocks and lower limbs. All others are described in HSP.

378: A. Myocarditis

Explanation: Clinical criteria for Kawasaki disease

> Fever of at least five days' duration.
>
> Presence of four of the following five principal features:
>
> 1. Changes in extremities;
> 2. Polymorphous exanthema;
> 3. Bilateral conjuctival injection;
> 4. Changes of the lips and oral cavity; and
> 5. Cervical lymphadenopathy.

> Exclusion of other diseases causing similar findings.

379: B. IVIG

Explanation: One-liner

380: C. 15-25%

Explanation: CAA develop in upto 25% of untreated patients in the 2nd to 3rd wk of illness. Giant coronary artery aneurysms (classic definition of >8 mm internal diameter)pose the greatest risk for rupture, thrombosis or stenosis, andmyocardial infarction.

381: D. Uveitis in most patients

Explanation: **sJIA**is characterized by arthritis, fever, rash, and prominent visceralinvolvement, including hepatosplenomegaly, lymphadenopathy, andserositis (pericarditis). The characteristic fever, defined as spiking temperaturesto ≥39°C (102.2°F), occurs on a daily or twice-daily basis forat least 2 wk, with a rapid return to normal or subnormal temperatures. The fever is often present in the evening and is frequentlyaccompanied by a characteristic faint, erythematous, macular rash. Theevanescent **salmon-colored lesions,** classic for sJIA, are linear or circularand are most commonly distributed over the trunk and proximalextremities. The classic rash is nonpruritic and migratorywith lesions lasting <1 hr. **Koebner phenomenon,** a cutaneous hypersensitivityin which classic lesions are brought on by superficial trauma,is often present. Heat can also evoke rash. Fever, rash, hepatosplenomegaly,and lymphadenopathy are present in >70% of affected children.

382: C. Uveitis is common

Explanation: Most cases of soJIA are ANA and RF negative.

383: A. Henoch-schonlein purpura

Explanation: Self-explanatory

384: C. Seizures

Explanation: Henoch-Schönlein purpura (HSP) is the most common small vesselvasculitis in childhood. It is characterized by a purpuric rash and commonlyaccompanied by arthritis and abdominal pain. Approximately 50% of patients with HSP develop renal manifestations,which vary from asymptomatic microscopic hematuria tosevere, progressive glomerulonephritis.

385: C. IgA

Explanation: In HSP, immunofluorescenceidentifies IgA deposition in walls of small vessels, accompaniedto a lesser extent by deposition of C3, fibrin, and IgM.

386: B. Pantoeaagglomerans

Explanation: A diagnosis of plant thorn synovitis should be considered in childrenwith monoarticular arthritis nonresponsive to anti-inflammatory therapy. Acute or chronic arthritis can occur after a plant thorn orother foreign object penetrates a joint. Unlike septic arthritis, childrenwith plant thorn synovitis are commonly afebrile. The most commonorganism seen with plant thorn synovitis is Pantoeaagglomerans,although cultures are often negative.

387: B. Morphea

Explanation: Morphea, also called localised scleroderma, is an idiopathic chronic fibrosing disorder manifesting as fibrosis and induration of skin and subcutaneous tissues. It is the most common type of scleroderma in childhood.

388: D. Within 6 months

Explanation: Renal disease can develop up to 6 months after diagnosis of HSP

389: B. XLD

Explanation: **Blau syndrome** is an autosomal dominant, familial form of sarcoidosisand is typified by the early onset of granulomatous inflammationinvolving the skin, eyes, and joints. Missense mutations in the CARD15/NOD2 gene on chromosome 16 have been found in affected familymembers and appear to be associated with development of sarcoidosis.

390: A. Fever lasting more than 4 weeks

Explanation: In KD, fever is characteristically high (≥38.3°C [101°F]), unremitting, andunresponsive to antibiotics. The duration of fever without treatment isgenerally 1-2 wk, but may persist for 3-4 wk. Periungual desquamation of the fingers and toesbegins 2-3 wk after the onset of illness.

391. **To monitor iron overload in long standing thalassemias, which is the best investigation?**
A. Liver iron measurement by MRI
B. Cardiac iron measurement by MRI
C. Serum Free iron measurment
D. Bone marrow ferritin levels after BM aspiration

392. **A 3year old male child was brought to OPD with complaints of short stature and poor growth. On examination, you noticed multiple brown Café-au-lait spots on the body & bony abnormalities involving the radial bone & thumb. Which among the following investigations will NOT be helpful in diagnosis?**
A. Chromosomal breakage test
B. Complementation test
C. Gene sequencing
D. DKC-1 gene detection

393. **One of your patients recently underwent splenectomy due to an autosomal recessive glycolytic enzyme deficiency. Which of the following is a common clinical sign in neonatal presentation of this disorder?**
A. Sepsis
B. Jaundice
C. Hepatomegaly
D. Hypertonia

394. **An 18-month-old toddler has microcytic anemia. Which dietary history finding best explains this?**
A. Lack of fresh vegetables in the diet
B. Intake of inadequate amounts of fruit juice
C. Intake of excessive amounts of vitamin C
D. Intake of large amounts of unmodified cow's milk

395. **Congenital cause of hypercoagulable states includes all except?**
A. Protein C deficiency
B. Protein S deficiency
C. MTHFR mutation
D. Lupus anticoagulant

396. **Mode of inheritance of congenital sideroblastic anemia is?**
A. Sporadic
B. X-linked recessive
C. Autosomal dominant
D. Autosomal recessive

397. **Kostmann's syndrome-treatment is :**
A. Anti-thymocyte globulin+ cyclosporin
B. Anti- thymocyte globulin + cycloporin +GM-CSF
C. G-CSF
D. GM- CSF

398. **Treatment of choice in Diamond-Blackfananemia is?**
A. IVIG
B. Erythropoietin
C. Corticosteroids
D. Azathioprine

399. **A child suffering from iron deficiency anemia was started on oral iron therapy. When will the peak reticulocytosis anticipated after starting iron?**
A. 2-3 days
B. 3-5 days
C. 5-7 days
D. 10-14 days

400. **A male child with Fanconi syndrome with nephrocalcinosis has a variant of dent disease. All are true except?**
A. Hypercalciuria
B. Proteinuria
C. Similar presentation in father
D. Rickets

401. **Which is Vitamin K dependent clotting factor?**
A. Factor VII
B. Factor I
C. Factor XI
D. Factor XII

402. **A 9-year-old boy comes with elevation of both PT and APTT. What is the diagnosis?**
A. Defect in extrinsic pathway
B. Defect in Intrinsic pathway
C. Platelet function defect
D. Defect in common pathway

403. **Neonate with blood group incompatibility not causing clinical hemolysis is?**
A. Bombay blood group
B. Kelly
C. Duff
D. Lewis

404. **A12-year-old child having microcytic hypochromic anemiahas blood picture showing Hb >40 gm/l, serum iron levels 20, transferrin saturation 40%,ferritin >800.The likely diagnosis is?**
A. Iron deficiency anemia
B. Hemochromatosis
C. Atransferrinemia
D. DMT1 gene mutation

405. **The type of disease having severe thrombocytopenia with decreased vWF and mildly decreased factor 8 levels in blood is?**
A. Type 1
B. Type 2A
C. Type 3
D. Pseudo-vWD

406. **In a child with Hemophilia A, the joint earliest affected in life by hemarthrosis is:**
A. Knee
B. Ankle
C. Hip
D. Elbow

407. **The most common presentation of G6PD deficiency is:**
A. Acute hemolytic crisis
B. Gallstones
C. Progressive pallor
D. Asymptomatic

391. a	392. d	393. b	394. d	395. d	396. b	397. c	398. c	399. c	400. c
401. a	402. d	403. d	404. c	405. d	406. b	407. d			

408. A 7 year old child had acute viral illness, followed by sudden appearance of non-palpable petechiae and purpura on the body. There was mild splenomegaly but no hepatomegaly. On investigations, Platelet count was 80000/mm³. CRP and TLC were elevated. Presence of which among the following rules out ITP as the likely cause?

A. Non-palpable lesions
B. Elevation of CRP
C. Age of the child
D. Presence of splenomegaly

409. What shall be the most common haematological abnormality seen in the child shown in picture?

A. Aplastic anemia
B. Thrombocytopenia only
C. JMML
D. PNH

410. Transient myeloproliferative disorder of newborn is seen in association with-

A. Turner syndrome
B. Down's syndrome
C. Neurofibromatosis
D. Ataxia telangectasia

411. The most common physical manifestation of Fanconi anemia is?

A. Hyperpigmentation
B. Short stature
C. Ear anomalies
D. Hypogonadism

412. What is the therapeutic dose of elemental iron in childhood iron-deficiency anemia?

A. 1-3 mg/kg/day
B. 3-6 mg/kg/day
C. 6-12 mg/kg/day
D. 12-15 mg/kg/day

413. The hematological abnormality seen in orotic aciduria is?

A. Microcytic anemia
B. Macrocytic anemia
C. Hemolysis
D. Thrombosis

414. What hematologic abnormality should you suspect in a newborn with bilateral absence of radii?

A. Thrombocytopenia
B. Anemia
C. Neutropenia
D. Pancytopenia

415. The following may present ashemolytic disease of the newborn:

A. Hereditary spherocytosis
B. Sickle cell trait
C. Dubin Johnson syndrome
D. Rotor syndrome

416. A 9-year-old African-American child is brought to the energy room complaining of painful swelling in his hands and feet, as well as abdominal pain. He is feverish [39°C (102.2°F)] and tachycardiac (140 beats per minute). He has not been exposed to any known infectious diseases. His eyes appear icteric, and his liver is enlarged. Of the choices below, which is a likely diagnosis?

A. Sickle cell disease
B. Acute lymphocytic leukemia
C. Henoch-Schonlein purpura
D. Aplastic anemia

417. Decreased LAP score is found in?

A. Leukaemoid reaction
B. Myelofibrosis
C. Pernicious anemia
D. Polycythemia

418. Non-hematologic manifestations of iron-deficiency in children include all except?

A. Cognitive defects
B. ADHD
C. Pagophagia
D. Pica

419. Not true regarding Rogers syndrome?

A. Thiamine responsive megaloblastic anemia
B. Conductive deafness
C. Diabetes mellitus
D. Autosomal recessive inheritance

420. What is characteristic of Gray platelet syndrome?

A. Absence of α granules
B. Absence of δ granules
C. Abnormal platelet aggregation in response to ristocetin
D. Pre-malignant condition

421. All of the following are true of childhood ITP except-

A. Prolonged clotting time
B. Preceded by viral infection
C. Increased bone marrow megakaryocytes
D. IVIG useful in severe cases

422. A 15-year old female presented to the emergency dept. with history of recurrent epistaxis, hematuria and hematochezia. There was a history of profuse bleeding from the umbilicus stump at birth. Previous investigations revealed normal PT, APTT, TT and fibrinogen levels. Her platelet counts as well as platelet function tests were normal but urea clot lysis test was positive. Which one of the following clotting factor is most likely to be deficient?

A. Factor X
B. Factor XI
C. Factor XII
D. Factor XIII

408. d	409. b	410. b	411. a	412. b	413. b	414. a	415. a	416. a	417. c
418. b	419. b	420. a	421. a	422. d					

423. Deferiprone use as chelator therapy needs monitoring of?

A. LFT
B. RFT
C. Blood cell counts
D. LDH

424. Hair on end appearance is classically seen in X-ray skull in?

A. Thalassemia
B. Hydrocephalus
C. Chronic malaria
D. Myelopthisis

425. In long standing thalassemia, the most common cause of death is?

A. BM suppression
B. Secondary infections
C. Cardiac causes
D. Malignancy

426. Microcytic anemia is seen in all of the following except?

A. Thalassemia
B. Sideroblastic anemia
C. Lead poisoning
D. Aplastic anemia

427. Transmission of G6-PD is?

A. X-linked
B. Acquired
C. AR
D. AD

428. Treatment useful to control mucosal bleeding in all types of vWD?

A. Desmopressin
B. FFP
C. Antifibrinolytics
D. vWF concentrates

429. Most common cause of acquired red cell aplasia in children is?

A. Parvovirus B19
B. Mitochondrial defects
C. Transient erythroblastopenia of childhood
D. HLH syndrome

430. RBCs replaced each day in a normal post-infant person is?

A. <1%
B. 1-5%
C. 5-10%
D. >10%

423. c	424. a	425. c	426. d	427. a	428. c	429. c	430. a

391: A. Liver iron measurement by MRI

Explanation: Quantitative liver iron measurement by approved MRI technology is the best indicator of total-body iron stores and should be obtained in chronically transfused patients after chronic transfusion therapy has initiated. The liver iron results will help guide the chelation regimen in thalassemias.

392: D. DKC-1 gene detection

Explanation: The child has features suggestive of Fanconi anemia. Diagnostic modalities include:

1. Chromosomal breakage test (in response to mitomycin C or diepoxybutane)
2. Complementation analysis (flow cytometric analysis of G_2 arrest in melphalan-exposed cells after transduction with retroviral vectors expressing normal fanconi anemia
3. Gene sequencing

393: B. Hepatomegaly

Explanation:

- Pyruvate kinase deficiency causes a congenital non-spherocytic hemolyticanemia with resultant hyperbilirubinemia.
- The breakdown may be so severe as to put the neonate at risk for bilirubin encephalopathy and require exchange transfusion.
- Blood transfusion may be indicated for the anemia. Splenectomy improves the anemia but does not cure the underlying defect. Sepsis is not a common initial presentation.
- Gallstones may occur causing hepatomegaly in those with pyruvate kinase deficiency.
- However, this typically occurs in adults rather than neonates. Seizures and hypertonia are not associated.

394: D. Intake of large amounts of unmodified cow's milk

Explanation:

- Inadequate dietary iron is the leading cause of iron deficiency anemia in children.
- Milk has low iron content, and the iron in cow's milk is not well absorbed. If a large percentage of dietary calories come from milk, the diet is apt to be low in iron.
- Additionally, microscopic gastrointestinal blood loss associated with the intake of unmodified cow's milk can contribute to iron deficiency.

395: D. Lupus anticoagulant

Explanation:

Risk Factors for Thrombosis	
Factor V Leiden	Venous
Homocystinuria	Venous and Arterial
Prothrombin G20210A	Inherited
Dysfibrinogenemia	Inherited
Antithrombin deficiency	
Protein C deficiency	
Mixed (Inherited and acquired)	
Protein S deficiency	
Hyperhomocysteinemia	
Elevated FVIII	
Acquired	
Acquired	
Malignancy	
Age	
Antiphospholipid antibody syndrome	
Previous thrombosis	
Hormonal therapy	
Immobilization	
Polycythemiavera	
Major surgery	
Essential thrombocythemia	
Pregnancy & puerperium	
Paroxysmal nocturnal hemoglobinuria	
Hospitalization	
Thrombotic thrombocytopenic purpura	
Obesity	
Heparin-induced thrombocytopenia	
Infection	
Disseminated intravascular coagulation	
APC resistance, nongenetic	
Unknown	
Elevated factor II, IX, XI	
Elevated TAFI levels	
Low levels of TFPI	

396: B. X-linked recessive

Explanation: Congenital sideroblastic anemia is usually an X-linked recessive disorder and is most commonly a result of mutations in erythrocytic isozyme 5-aminolevulinic acid synthetase, the rate-limiting enzyme reaction in heme synthesis

397: C. G-CSF

Explanation:

- Kostmann syndrome, also known as severe congenital neutropenia, autosomal recessive type 3 (SCN3) and Kostmann disease, is a rare autosomal recessive form of severe chronic neutropenia usually detected soon after birth.
- An absolute neutrophil count (ANC) chronically less than 500/mm3 is the main sign of Kostmann's.
- A standard bone marrow test can give correct diagnosis.Bone marrow usually shows the

presence of early granulocytes (promyelocyte/myelocyte arrest) but few maturing forms are seen; neutrophil survival is normal.

- Homozygous mutations in the HAX1 gene are associated with Kostmann disease, i.e. the "classical," autosomal recessive form of severe congenital neutropenia (SCN3).
- Regular administration of exogenous granulocyte colony-stimulating factor (Filgrastim) clinically improves neutrophil counts and immune function and is the current mainstay of therapy, although this may increase risk for myelofibrosis and acute myeloid leukemia in the long term

398: C. Corticosteroids

Explanation: Corticosteroids are a mainstay of therapy in Diamond-Blackfananemia and approximately 80% of patients initially respond. Patients who are nonresponders or who fail to tolerate corticosteroid therapy require transfusions at intervals of 3-5 wk to maintain a haemoglobin level greater than 8 g/dL.Hematopoietic stem cell transplantation (HSCT) can be curative.

399: C. 5-7 days

Explanation:

Time after iron administration	Response
12-24 hr	Replacement of intracellular iron enzymes; subjective improvement; decreased irritability; increased appetite
36-48 hr	Initial bone marrrow response;
48-72 hr	Reticulocytosis, peaking at 5-7 days
4-30 days	Increase in hemoglobin level
1-3 mo	Repletion of stores

400: C. Similar presentation in father

Explanation:

- Dent's disease (or Dent disease) is a rare X-linked recessive inherited condition that affects the proximal renal tubules of the kidney.
- It is one cause of Fanconi syndrome, and is characterized by tubular proteinuria, hypercalciuria, calcium nephrolithiasis, nephrocalcinosis and chronic renal failure.

401: A. Factor VII

Explanation: Vitamin K–dependent carboxylation of Glu to Gla residues occurs in the amino terminal regions of factors II, VII, IX, and X and also proteins C and S

402: D. Defect in common pathway

Explanation:

Hemostatic Disorders and Coagulation Test Abnormalities
Prolonged activated partial thromboplastin time (aPTT)
No clinical bleeding – decreased factors XII, high-molecular-weight kininogen, protein kinase
Variable, but usually mild, bleeding – decreases factor XI, mild decrease FVIII and FIX
Frequent, severe bleeding – severe deficiencies of FVIII and FIX
Heparin
Prolonged prothrombin time (PT)
Factor VII deficiency(extrinsic factors)
Vitamin K deficiency – early
Warfarin anticoagulation
Prolonged aPTT and PT
Factor II, V or X deficiency(common pathway factors)
Vitamin K deficiency – late
Direct thrombin inhibitors
Prolonged thrombin time
Heparin or heparin-like inhibitors
Mild or no bleeding – dysfibrinogenemia
Frequent, severe bleeding – afibrinogenemia
Prolonged PT and/or aPTT not correct with mixing with normal plasma
Bleeding – specific factor inhibitor
No symptoms, or clotting and/or pregnancy loss – lupus anticoagulant
Disseminated intravascular coagulation
Heparin or direct thrombin inhibitor
Abnormal clot solubility
Factor XIII deficiency
Inhibitors or defective cross-linking
Rapid clot lysis
Deficiency of alpha$_2$-antiplasmin or plasminogen activator inhibitor 1
Treatment with fibrinolytic therapy

403: D. Duff

Explanation: Lewis-anti Lewis incompatibility reaction does not produce clinical hemolysis in the newborn.

404: C. Atransferrinemia

Explanation: Atransferrinemia is characterised by microcytic hypochromic picture, low serum iron and low TIBC.Transferrin is low leading to high ferritin. Hemochromatosis typically doesn't have MCHC or low iron.

405: D. Pseudo-vWD

Explanation: Platelet-type, pseudo-VWD occurs when a mutation in platelet GPIb causes spontaneous binding to VWF and also presents with decreased VWF activity, loss of high-molecular-weight multimers, and thrombocytopenia similar to type 2B VWD. Specific testing is required to distinguish the 2 conditions.

406: B. Ankle

Explanation: Although bleeding may occur in any area of the body, the hallmark of hemophilic bleeding is hemarthrosis. Bleeding into the joints may be induced by minor trauma; many hemarthroses are spontaneous. The earliest joint hemorrhages appear most commonly in the ankle. In the older child and adolescent, hemarthroses of the knees and elbows are also common.

407: D. Asymptomatic

Explanation: Most individuals with G6PD deficiency are asymptomatic, with no clinical manifestations

of illness unless triggered by infection, drugs, or ingestion of fava beans. Typically, hemolysis ensues in about 24-48 hr after a patient has ingested a substance with oxidant properties. Insevere cases, hemoglobinuria and jaundice result, and the haemoglobin concentration may fall precipitously.

408: D. Presence of splenomegaly

Explanation: Presence of splenomegaly and significant lymphadenopathy rule out ITP as the cause of skin or mucosal bleeds in children.

409: B. Thrombocytopenia only

Explanation: This is a child with absent radial bone (radial forearm defect), most likely due to TAR syndrome (Thrombocytopenia with absent radii syndrome).

Note the presence of shortened/hypoplastic forearm with thumb being present (Fanconi anemia has absent thumb as well).

Points of difference of TAR syndrome V/S Fanconi anemia (from MCQ perspective)

- In Fanconi anemia, thrombocytopenia is rarely present at birth; it develops later on in childhood or even in adulthood.
- Radial defects are seen in only 30% of patients with Fanconi anemia; when present, such defects are usually associated with absent thumb, unlike in TAR, where radial defects are the sine qua non and the thumb is usually present, even though it could be abnormal

410: B. Down's syndrome

Explanation: Neonates with Down syndrome (DS) have a unique predilection to develop transient myeloproliferative disorder (TMD), a rare clonal myeloproliferation characterized by peripheral leukocytosis indistinguishable at presentation from acute megakaryocytic leukemia, FAB M7, or acute myeloid leukemia (AML) with minimal differentiation, FAB M0. Its unique characteristics include a relative paucity of leukemic blasts within the marrow, variable pancytopenia, a propensity for mild to life-threatening hepatic infiltration, and typically a spontaneous regression without any intervention.

411: A. Hyperpigmentation

Explanation:

Charactoristic Physical Anomalios in Fanconi Anemia	
Anomaly	Approximate frequency (% of patients)
Skin pigment changes ± lait spots	55
Short stature	51
Upper limb abnormalities (thumbs, hands, radii, ulnas)	43
Hypogonadal and genital changes (mostly male)	35
Other skeletal findings (head/face, neck, spine)	30
Eye/lid/epicanthal fold anomalies	23
Renal malformations	21
Gastrointestinal / cardiopulmonary malformations	11
Hip, leg, foot, toe abnormalities	10
Ear anomalies (external and internal, defness	9

412: B. 3-6 mg/kg/day

Explanation: The therapeutic dose should be calculated in terms of elemental iron. A daily total dose of 3-6 mg/kg of elemental iron in 3 divided doses is adequate, with the higher dose used in more severe cases. The maximum dose would be 150-200 mg of elemental iron daily.

413: B. Macrocytic anemia

Explanation: Orotic aciduria is a rare autosomal recessive disorder that usually appears in the 1st yr of life and is characterized by growth failure, developmental retardation, megaloblastic anemia, and increased urinary excretion of orotic acid.

414: A. Thrombocytopenia

Explanation:

- Thrombocytopenia with absent radius (TAR) syndrome is one of the congenital thrombocytopenia syndromes.
- The thrombocytopenia severity usually lessens with advancing age. Patients may occasionally have other congenital abnormalities.
- Wiskott-Aldrich syndrome (WAS) is another congenital thrombocytopenia syndrome in which boys are affected with thrombocytopenia, eczema, and severe immunodeficiencies.
- Congenital amegakaryocytic thrombocytopenia usually presents in the first weeks of life with petechiae and purpura.

415: A. Hereditary spherocytosis

Explanation: CAUSES OF HAEMOLYTIC DISEASE OF THE NEWBORN

- Immune: (Rh, ABO, other)
- Membrane defects: Spherocytosis, elliptocytosis
- Enzyme defects: G6PD, PK, hexokinase
- Sepsis
- Polycythemia: IDM, fetal transfusion.

416: A. Sickle cell disease

Explanation: Children with sickle cell disease often present with painful swelling of the hands and feet, as well as pain in the abdomen and back. These children are often asymptomatic in early months of life.

417: C. Pernicious anemia

Explanation: Leukocyte alkaline phosphatase is a test how much alkaline phosphatase (ALP) is inside white blood cells.

A. Higher-than-normal results may be due to:
   a. Leukemoid (leukemia-like) reaction.
   b. Myelofibrosis.
   c. Polycythemiavera.

B. Lower-than-normal results may be due to :
   a. Aplastic anemia
   b. Chronic granulocytic leukemia
   c. Pernicious anemia.

418: B. ADHD

Explanation: Nonhematologic systemic effects of iron deficiency

- Both iron deficiency and iron-deficiency anemia are associated with impaired neurocognitive function in infancy. There is also an association of iron-deficiency anemia and later, possibly irreversible, cognitive defects.
- Some studies suggest an increased risk of seizures, strokes, breathholding spells in children, and exacerbations of restless leg syndrome in adults.
- Other nonhematologic consequences of iron deficiency include pica, the desire to ingest nonnutritive substances, and pagophagia, the desire to ingest ice. The pica can result in the ingestion of lead-containing substances and result in concomitant plumbism.

419: B. Conductive deafness

Explanation: **Thiamine-responsive megaloblastic anemia** (Rogers syndrome) is a very rare autosomal recessive disorder characterized by megaloblasticanemia, sensorineural deafness, and diabetes mellitus. Congenitalheart defects, arrhythmias, visual problems, short stature, trilineagemyelodysplasia, and strokes are also described. Thiamine-responsivemegaloblastic anemia usually presents in infancy but may occasionally develop in childhood and adolescence and occurs in several ethnicallydistinct populations. The bone marrow is characterized not only by megaloblastic changes but also by ringed sideroblasts. The defect iscaused by mutations in the SCL19A2 gene on chromosome 1, which encodes a high-affinity plasma membrane thiamine transporter. Continuous thiamine supplementation usually reverses the anemia and diabetes but not existing hearing defects.

420: A. Absence of α granules

Explanation: **Gray platelet syndrome** is caused by the absence of platelet α granules, resulting in large platelets that are large and appear gray on Wright stain of peripheral blood. In this rare syndrome, aggregation and release are absent with most agonists other than thrombin and ristocetin. Electron microscopic studies are diagnostic.

421: A. Prolonged clotting time

Explanation: Bleeding time is affected in ITP, not clotting time.

422: D. Factor XIII

Explanation: Screening tests for factor XIIIdeficiency are based on the observation that there is increased solubilityof the clot because of the failure of crosslinking. The normal clotremains insoluble in the presence of 5M urea, whereas in a patientwith XIII deficiency, the clot dissolves.

423: C. Blood cell counts

Explanation: Deferiprone, a small molecule, effectively enters cardiac tissue and maybe more effective than other chelators in reducing cardiac hemosiderosis.The most serious side effect of deferiprone is transient agranulocytosis,which occurs in 1% of patients. It is associated with rare deathswhere patients were not adequately monitored. The use of deferipronerequires weekly white blood cell counts.

424: A. Thalassemia

Explanation: **Hair** on **end appearance** of the skull is a characteristic feature of chronic haemolysis usually seen in patients with thalassaemia and sickle cell anaemia. It results from accentuated vertical trabeculae between the inner and outer tables of the skull because of excessive bone marrow hyperplasia.

425: C. Cardiac causes

Explanation: Cardiac disease is the major cause of death in thalassemia.Serial echocardiograms should be monitored to evaluate cardiacfunction and pulmonary artery pressure. Pulmonary hypertension frequently occurs in non-transfused thalassemia patients and may be anindication for transfusion therapy.

426: D. Aplastic anemia

Explanation: Causes of microcytic anemia

Children and adolescents

Iron deficiency anemia
Thalassemia trait
Other hemoglobinopathies
Lead toxicity
Chronic inflammation
Sideroblastic anemia

427: A. X-linked

Explanation: One-liner

428: C. Antifibrinolytics

Explanation: Treatment of vWD

Treatment	VWD Types
Desmopressin	Type 1 VWD Some type 2 VWD (use with caution)
von Willebrand factor concentrates	Type 3 VWD Type 2 VWD Severe type 1 VWD (or type 1 clearance defects)
Antifibrinolytics	Mucosal bleeding, all types or VED

429: C. Transient erythroblastopenia of childhood

Explanation: Transient erythroblastopenia of childhood (TEC) is the most commonacquired red cell aplasia occurring in children. It is more prevalent thancongenital hypoplastic (Diamond-Blackfan) anemia. This syndrome ofsevere, transient hypoplastic anemia occurs mainly in previouslyhealthy children between 6 months and 3 yr of age; most of the childrenare older than 12 months at onset.

430: A. <1%

Explanation: Normal RBC survival time is 110-120 days (half-life: 55-60 days), and thus, approximately 0.85% of the most senescent RBCs are removed and replaced each day.

431. A 2 year old child with large untreated boil on right thigh, comes in septic shock. He is started on IV fluids & after giving 3 NS boluses, he develops basal crepitations but the shock continues. He is also administered a dose of Hydrocortisone & started on Dopamine infusion. Currently he is on Dopamine infusion @ 20 micrograms/kg/min. What is the next therapy to consider?

A. Start 5% Albumin infusion
B. Start I.V. Antibiotics
C. Add epinephrine infusion i.v.
D. Add norepinephrine infusion i.v.

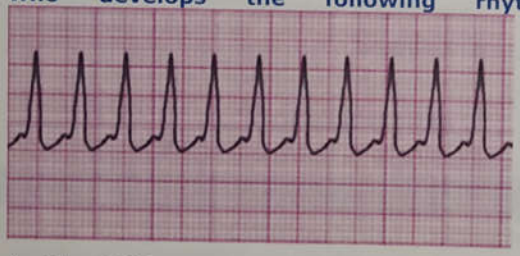
432. In an oliguric child needing intravenous fluids, insensible losses should be replenished with which fluid?

A. 0.9% saline
B. 0.45% saline
C. 0.45% saline in 5% dextrose
D. 0.9% saline in 10% dextrose

433. Most common type of shock in children?

A. Septic
B. Hypovolemic
C. Neurogenic
D. Cardiogenic

434. What is the immediate therapy of choice who develops the following rhythm?



A. Place ECD
B. Give amiodarone
C. Give adenosine
D. Wait and watch

435. A 2 year old child has severe dehydration and unrecordable BP. Attempts to obtain intravenous access fail. What is the next step in management?

A. Keep trying for half hour
B. Do venesection
C. Obtain intraosseous access
D. Do IJV catheterization

436. Most common bone used for intraosseous access?

A. Tibia
B. Fibula
C. Humerus
D. Sternum

437. In a child with cardio-pulmonary arrest, intraosseous access should be attempted if venous access is not available within?

A. 1 min
B. 2 min
C. 3 min
D. 5 min

438. The confirmatory method for correct endotracheal tube placement in a child is?

A. Chest X ray
B. Capnography
C. Impedance technique
D. Mist formation in tube

439. Hypotension in a 5 month child is defined as SBP below?

A. 60 mmHg
B. 70 mm Hg
C. 80 mm Hg
D. 90 mm Hg

440. A 3year old child presented with HR of 122/min, capillary refill time of 4 seconds and cold peripheries with BP of 86/40 mm Hg. What is the next step in management?

A. RL bolus @ 100 ml/kg over 6 hr
B. 0.45% NS @ 10 ml/kg
C. 0.9% NS@ 20 ml/kg
D. 5% Albumin infusion @ 10ml/kg

441. Which among the following is incorrect regarding shock in children?

A. Hypovolemia is a common cause
B. Septic shock may show bounding pulses with warm peripheries
C. Tachycardia is often present
D. Fall in BP is an early sign

442. Treatment of choice in high-altitude pulmonary edema in a child?

A. Supplemental O2
B. Dexamethasone
C. Frusemide
D. Acetazolamide

443. Cricothyroidotomy is preferred over endotracheal intubation in children in?

A. Cleft palate
B. Acute epiglottitis
C. Complete airway obstruction
D. Tracheomalacia

444. Dose of dopamine in Paediatric shock management?

A. 1-2 μg/kg/min
B. 2-10 μg/kg/min
C. 10-20 μg/kg/min
D. 20-40 μg/kg/min

445. The hallmark finding in traumatic brain injury (TBI) in a child is?

A. Pupil abnormalities
B. Coma
C. Brisk reflexes
D. Papilledema

446. Best way to monitor shock in children is?

A. Heart rate
B. Urine output
C. Ejection fraction
D. Preload

447. Which among the following inotropic agents is contraindicated in cardiogenic shock?

A. Amrinone
B. Dopamine
C. Epinephrine
D. Norepinephrine

431. d	432. c	433. b	434. c	435. c	436. a	437. a	438. b	439. b	440. c
441. d	442. a	443. c	444. c	445. b	446. d	447. d			

448. **A 6 year old boy developed steroid resistant nephrotic syndrome. He was diagnosed as FSGS on renal biopsy and started on oral cyclosporine @ 3 mg/kg/day. Two days later, he developed ataxia, slurred speech, nystagmus, hypertension and altered sensorium. MRI brain was suggestive of hyperintense lesions and some demyelination in parieto-occipital regions on T2 images. Which among the following best describes the reason for this presentation?**

A. PRES
B. Multiple sclerosis
C. GBS
D. Pseudotumor cerebri

449. **Which among the following are seen in early raised intracranial pressure in children?**

A. Apnea and Bradypnea
B. Apnea and Tachypnea
C. Tachypnea and Bradypnea
D. Tachypnea and Hyperpnea

450. **Heliox is useful in the emergency management of all of the following conditions except?**

A. Acute laryngotracheobronchitis
B. Vascular ring
C. Subglottic stenosis
D. Acute bronchiolitis

448. a	449. d	450. d

431: D. Add norepinephrine infusion i.v.

Explanation: This is a child with septic shock. The shock is resistant to fluids & Dopamine. The next best step will be to add IV norepinephrine in this patient.

Always remember:

- Any shock, start with IV fluids; Normal saline preferred; max 3 doses of 20 ml/kg each
- Move to dopamine @ 10-20 microgram per kg per min
- If still in shock, Add Epinephrine in cold shock & Add Norepinephrine in warm/septic shock
- Hydrocortisone & IV antibiotics additionally in septic shock

432: C. 0.45% saline in 5% dextrose

Explanation:

Adjusting Fluid Therapy for Altered Renal Output
Oliguria / Anuria
Replacement of insensible fluid losses (25-40% of maintenance) with D5 $^1/_2$ NS Replace urince output mL/mL with D5 $^1/_2$ NS ± KCl
Polyiria
Replacement of insensible fluid losses (25-40% of maintenance) with D5 $^1/_2$ NS ± KCl
Measure urine electrolytes
Replace urine output mL/mL with solution based on measured urine electrolytes

433: B. Hypovolemic

Explanation:

- MC type of shock in children overall: Hypovolemic
- MC type of shock in neonates: Septic

434: C. Give adenosine

Explanation: This is SVT. For acute attack, Adenosine I.V. is the therapy of choice

435: C. Obtain intraosseous access

Explanation: IO cannulation is recommended for patients for whom IV access proves difficult or unattainable, even in older children. For children in shock and poor venous access, IO cannulation can be life-saving. Anterior aspect of tibia is the preferred site.

436: A. Tibia

Explanation: Refer to Q.5 explanation

437: A. 1 min

Explanation: If venous access is not available within 1 min in a child with cardiopulmonary arrest, an IO needle should be placed in the anterior proximal tibia (with care taken to avoid traversing the epiphyseal plate).

438: B. Capnography

Explanation: Detection of end-tidal CO2 measurement using capnography is the management technique of choice to detect correct endotracheal tube placement.

439: B. 70 mm Hg

Explanation: Hypotension in children is defined as a systolic blood pressure:

- Below 60 mm Hg in term neonates (0 to 28 days)
- Below 70 mm Hg in infants (1 month to 12 months)
- Below 70 mm Hg + (2 x age in years) in children 1 to 10 years
- Below 90 mm Hg in children above 10 years of age

440: C. 0.9% NS@ 20 ml/kg

Explanation: According to Nelson 20th Ed and PALs guidelines, isotonic saline bolus @ 20 ml/kg is the first step in shock management

Initial resuscitation: Push boluses of 20 cc/kg isotonic saline or colloid up to and over 60 cc/kg until perfusion improves or unless rales or hepatomegaly develop.

Correct hypoglycemia and hypocallcemia. Being antibiotics.

441: D. Fall in BP is an early sign

Explanation: Hypotension is a late finding in shock and indicates significant decompensation has already taken place.

442: A. Supplemental O2

Explanation: Descent with supplemental oxygen is the treatment of choice for HAPE in children. When feasible, or in the absence of medical care, rapid descent of at least 500-1,000 m usually results in rapid recovery. As withall altitude illness the magnitude of the descent is tailored to the resolution of symptoms. Oxygen and bed rest without descent can be safe and effective treatment for mild HAPE in children where careful medicalobservation is available.

443: C. Complete airway obstruction

Explanation: Experts generally agree that in settings of known complete airway obstruction, endotrachealintubation should be avoided, and emergency cricothyroidotomy performed instead.

444: C. 10-20 µg/kg/min

Explanation: Dopamine produces renal vasodilatation in low dose, i.e.1-10 µg/kg/min and shock dose is 10-20 µg/kg/min.

445: B. Coma

Explanation: The hallmark of **severe TBI** is coma (**GCS score 3-8**). Often, coma is seen immediately after the injury and is sustained. In some cases, such as with an epidural hematoma, a child may be alert on presentation but may deteriorate after a period of hours.

446: D. Preload

Explanation: In childhood shock, best way to monitor is Central venous pressure (CVP) monitoring, also called preload monitoring

447: D. Norepinephrine

Explanation: Even though they may be beneficial in other forms of shock, agents that improve blood pressure by increasing SVR, such as norepinephrine and vasopressin, should generally be avoided in patients with cardiogenic shock. These agents may cause further decompensation and potentially precipitate cardiac arrest as a result of the increased afterload and additional work imposed on the myocardium. Ref. Nelson 20th Ed.

448: A. PRES

Explanation: The child has developed PRES (Posterior reversible encephalopathy syndrome), which is a rare but known CNS complication with cyclosporine use.

- Presents with ataxia, nystagmus, hypertension and confused state
- Can develop even at therapeutic doses other than toxicity.
- MRI shows frontal and parieto-occipital hyperintensities and signal enhancement in T2 images. Some degree of demyelination may be present
- Treatment is supportive and patient generally recovers in most cases

449: D. Tachypnea and Hyperpnea

Explanation: Early rise in ICP results in stimulation of respiratory centers, leading to increases in the rate (tachypnea) and depth (hyperpnea) of respiration. The resultant decrease in Paco2 and elevation of cerebrospinal fluid pH lead to cerebral vasoconstriction and amelioration of intracranial hypertension.

450: D. Acute bronchiolitis

Explanation: Helium-oxygen mixture (heliox) is useful in overcoming airway obstruction and improving ventilation. Helium is much less dense and slightly more viscous than nitrogen. When substituted for nitrogen, helium helps maintain laminar flow across an obstructed airway, decreases airway resistance, and improves ventilation. It is especially helpful in diseases of large airway obstruction in which turbulent airflow is more common, such as acute laryngotracheobronchitis, subglottic stenosis, and vascular ring. It is also used in patients with severe status asthmaticus.

451. Daily fluid maintenance requirement in a 16 kg child is:

A. 1200 ml
B. 1300 ml
C. 1500 ml
D. 1600 ml

452. Regarding Infantile tremor syndrome(ITS), all are true except:

A. More common in males
B. Hyperpigmentation is seen
C. Fine tremors are seen
D. Cortical atrophy is seen

453. A female child aged 6 years presents to casualty with # pelvis & severe bleeding from the perineum post traumatic assault. What shall be the immediate step in management?

A. Inform police before starting treatment
B. Assess & secure airway
C. Start chest compression
D. Start blood transfusion

454. A patient is found to have optic gliomas and his examination shows multiple dark lesions as shown. What is the likely diagnosis?

A. NF-1
B. NF-2
C. Incontinentia pigmenti
D. Linear naevus syndrome

455. Which vaccine is contraindicated in child with history of convulsions?

A. DPT
B. Measles
C. Typhoid
D. BCG

456. "Dopamine hypothesis" has been postulated for which among the following neuropsychiatric disorders?

A. Rett Syndrome
B. Autism
C. Asperger disorder
D. ADHD

457. X-ray of choice for age detection in a 6 month old child is?

A. Skull
B. Hand
C. Knee
D. Shoulder

458. Not true regarding primary ciliary dyskinesia is/are?

A. May manifest in neonates
B. Autosomal dominant inheritance in most cases
C. Both males and females may show infertility
D. Bronchiectasis is common

459. A 13month old child who is unimmunized, comes to OPD. Which among the following shall be given at the first visit? (Recent AIIMS MCQ)

A. BCG, Pentavalent vaccine, Measles, Hepatitis B
B. Pentavalent vaccine, Measles, Hepatitis B
C. BCG, DPT, OPV, Hepatitis B
D. Pentavalent vaccine, Rotavirus vaccine, Hepatitis B

460. Structure derived from umbilical artery?

A. Ligamentum arteriosum
B. Ligamentum venosum
C. Medial umbilical ligament
D. Ligamentum teres

461. Reduction or absence of which skin protein plays a pivotal role in the pathogenesis of childhood atopic dermatitis?

A. Dubin
B. Filaggrin
C. Desmoglein
D. Olein

462. Not a component of Pentalogy of Cantrell?

A. Omphalocele
B. Sternal cleft
C. Epigastric hernia
D. Ectopia cordis

463. Macrocephaly is seen in?

A. Metachromatic leukodystrophy
B. Adrenoleukodystrophy
C. Canavan's disease
D. Krabbe's disease

464. Which of these medicines taken during pregnancy is responsible for the defect seen in the child below?

A. Ferrous sulphate
B. Isotretinoin
C. Folic acid
D. Metoclopramide

451. b	452. c	453. a	454. a	455. a	456. d	457. d	458. b	459. b	460. c
461. b	462. c	463. c	464. b						

465. A child presented with anemia, kinky hair, intellectual disability and seizures. Which of the following is the most likely diagnosis?

A. Menkes disease
B. Down syndrome
C. Iron deficiency anemia
D. Lesch-nyhan syndrome

466. Which of the following best describes the correct sequence for eruption of permanent teeth?

A. Central incisors, first premolars, canines, second molars
B. First molars, central incisors, canines, first premolars
C. Central incisors, lateral incisors, canines, first molars
D. First molars, lateral incisors, canines, first premolars

467. A 5-yr-old girl is evaluated for severe mental retardation, microcephaly, hand-wringing movements, poor growth and weight gain, and seizures. Genetic analysis indicates a mutation in a transcription factor called MeCP2 that normally functions to silence transcription of numerous genes. This finding indicates that she has which of the following disorders?

A. Juvenile Huntington disease
B. Metachromatic leukodystrophy
C. Rett syndrome
D. Adrenoleukodystrophy

468. Not seen in Sturge Weber syndrome?

A. Port wine stain
B. Contralateral glaucoma
C. Ipsilateral leptomeningeal angiomas
D. Focal seizures

469. Which among the following is the therapy of choice for childhood eczema?

A. Topical calcipotriol
B. Topical corticosteroids
C. Systemic corticosteroids
D. Antihistamines

470. A 2-yr-old boy undergoing outpatient surgery for tonsillectomy vomits while recovering from general anesthesia in the postoperative recovery area. Within 1-2 min he develops tachypnea, chest retractions, and hypoxemia. Which of the following is the most likely explanation for these findings?

A. Acute blood loss from postoperative bleeding
B. Anesthetic reaction
C. Bacterial infection and toxin release
D. Acute airway obstruction from aspirated material

471. BCG vaccine, though not protective, provides effective protection against which among the following?

A. CNS and disseminated TB
B. Pulmonary TB
C. Pulmonary and skeletal TB
D. Pulmonary and CNS TB

472: Adrenal cortex of fetus maximally releases?

A. Cortisol
B. DHEAS
C. Progesterone
D. Corticosterone

473. Condition associated with more than 20% incidence of chromosomal anomalies?

A. Spina bifida
B. Gastroschisis
C. Omphalocele
D. Cleft palate

474. Not useful in the management of infantile hemangiomas?

A. Corticosteroids
B. Propranolol
C. Imiquimod
D. Indomethacin

475. All of the following produce microcephaly in a child, except?

A. Williams syndrome
B. Lissencephaly
C. Sturge Weber syndrome
D. Patau syndrome

476. Levine criteria have been proposed for the diagnosis of which among the following neurological conditions?

A. HIE
B. Cerebral palsy
C. Mitochondrial diseases
D. GBS

477. Most common route of spread of TB infection to a neonate born by caesarean section is?

A. Skin contact
B. Aspiration of infected amniotic fluid
C. Inhalational route
D. Hematogenous

478. Feature seen in pallid breath holding spells, in comparison to cyanotic breath holding spells is?

A. Pallid spell is more common
B. Due to sympathetic overactivity
C. Painful stimulus can abort this spell
D. Bradycardia is seen

479. Vaccine(s) which is/are recommended by Indian academy of Paediatrics (IAP) at 2 years of age is/are?

A. Varicella
B. MMR
C. Pneumococcal conjugate
D. Typhoid conjugate booster

480. Pharmacological therapy of choice for nocturnal enuresis?

A. Oral desmopressin
B. Nasal desmopressin
C. Oral imipramine
D. Oral oxybutynin

481. All are features of Refsum disease except:

A. Deficiency of alpha-hydroxylase
B. Defect in beta- oxidation
C. CSF protein concentration is elevated
D. Peripheral neuropathy

465. a	466. c	467. c	468. b	469. b	470. d	471. a	472. b	473. c	474. d
475. c	476. b	477. c	478.	479. d	480. a	481. b			

482. True regarding Minimal enteral nutrition (MEN) is all except:

A. Can be given even when baby is on ventilator
B. Can be given to hemodynamically unstable babies
C. Only expressed breast milk to be used
D. Contraindicated in suspected malrotation

483. A 4-year-old child is brought to OPD with complaints of eating mud, plaster of paris and paint flakes. Which among the following is TRUE regarding this child?

A. Not associated with iron deficient state
B. May be associated with Lead poisoning
C. Mental retardation seen in most cases
D. Positive family history is seen

484. Fever, headache, severe myalgias and a dark rash on hands and arms in a 7 yr old boy who went to his grandparents living in Chamba, Himachal pradesh. On examination, he has fever, petechial rash, hepatosplenomegaly and meningismus. Lab investigations show WBC of 3000/mm3, Platelet count of 90,000 and serum sodium 129. Which among the following is TRUE?

A. Man to man transmission seen
B. Skin rash is due to complement mediated damage
C. Bone marrow examination is needed
D. Azithromycin is a good alternative

485. Cause of death in Potter syndrome?

A. Anuria
B. CVS defects
C. Pulmonary hypoplasia
D. CNS defects

486. Not an indication for audiometric test in children?

A. Mechanical ventilation for 48 hours
B. Congenital CMV infection
C. Extreme prematurity
D. Acute pyogenic meningitis

487. Most common genetic cause of mental retardation?

A. Down syndrome
B. Fragile-X syndrome
C. Congenital hypothyroidism
D. Tuberous sclerosis

488. Identify the incorrect match for abnormal tongue and underlying diseases?

A. Strawberry tongue: Scarlet fever
B. Strawberry tongue: Kawasaki disease
C. Red beefy tongue: Vitamin B12 deficiency
D. Black hairy tongue: Hemolytic anemia

489. Inheritance of achondroplasia is?

A. Autosomal dominant
B. Autosomal recessive
C. X-linked recessive
D. Mitochondrial

490. Not seen in Reye syndrome?

A. Hypoglycemia
B. Raised AST
C. Raised serum bilirubin
D. Raised blood ammonia

491. Marfan syndrome is due to mutation in gene present on?

A. 11p
B. 11q
C. 15p
D. 15q

492. Most common neuro-behavioural disorder in children is?

A. ADHD
B. Autism
C. Dyslexia
D. Rett syndrome

493. Not consistent with a diagnosis of fulminant hepatic failure in children?

A. Biochemical evidence of acute liver injury lasting 12 weeks or less
B. PT >15 sec
C. INR >1.5
D. Hepatic encephalopathy

494. Severe growth restriction with cholestasis and iron overload is seen in?

A. Neuronal ceroid lipofuchinosis
B. GRACILE syndrome
C. Apert syndrome
D. Alagille syndrome

495. Children with atopic dermatitis are prone to skin colonization by which bacteria?

A. Pseudomonas
B. Staphylococcus aureus
C. Bacteroides
D. Streptococcus mutans

496. Which bacteria is most frequently associated with dental caries in children?

A. Staphylococcus epidermidis
B. HACEK organisms
C. Streptococcus bovis
D. Streptococcus mutans

497. Enuresis is defined as the repeated voiding of urine into clothes or bed at least twice a week for at least 3 consecutive months in a child who is at least?

A. 3 years age
B. 4 years age
C. 5 years age
D. 6 years age

498. Typically, children stop taking daytime naps at which of the following ages?

A. 18 months
B. 24 months
C. 36 months
D. 48 months

499. On physical examination, a newborn male has deficient abdominal musculature and undescended testes. You presumptively diagnose the child with which of the following?

A. VATER association
B. Cushing's triad
C. Potter's syndrome
D. Eagle-Barrett syndrome

500. Allergen immunotherapy for desensitization in a child is contraindicated in all except?

A. Vernal keratoconjunctivitis
B. Child on propranolol
C. ABPA
D. Autism

482. b	483. b	484. d	485. c	486. a	487. a	488. d	489. a	490. c	491. d
492. c	493. a	494. b	495. b	496. d	497. c	498. d	499. d	500. a	

451: B. 1300 ml

Explanation: Use Holliday-Segar method.
16 yr: 1000 ml + 50 x 6 = 1300 ml.

452: C. Fine tremors are seen

Explanation: m Coarse tremors are hallmark in Infantile tremor syndrome

453: B. Assess & secure airway

Explanation: **Remember:**

- In Nontraumatic CPR, sequence is CAB
- In traumatic CPR, sequence is ABC.
- Here, we shall assess & secure airway and then move to circulation management

454: A. NF-1

Explanation: The patient has multiple café-au-lait macules and optic nerve gliomas. This fulfills the criteria for Neurofibromatosis-1 (NF-1).

Diagnostic Criteria for Neurofibromatosis Type 1

Presence of two or more of the following features is considered diagnostic

- Six cafeaulait spots >5mm in diameter in prepubertal children or >15mm in postpubertal children
- Two or more neurofibromas or one plexiform neurofibroma
- Freckling in the axillary or inguinal region
- Optic glioma
- Two or more iris hamartomas (Lisch nodules)
- A distinctive osseous lesion (sphenoid dysplasia or thinning of long bones)
- A first - degree relative with neurofibromatosis type 1

455: A. DPT

Explanation: Contraindications of DPT(pertussis component):
Anaphylaxis, encephalopathy within 7 days of previous dose, Progressive / evolving neurological illness, excessive crying after vaccination

456: D. ADHD

Explanation: Children with ADHD have approximately a 5-10% reduction in the volume of these brain structures. Functional MRI findings suggest low blood flow to the striatum. Functional MRI data also suggest deficits in a widespread functional networks for selective and tonic attention in ADHD, that include the striatum, prefrontal regions, parietal lobe, and temporal lobe. The prefrontal cortex and basal ganglia are rich in dopamine receptors. This knowledge, plus data about the dopaminergic mechanisms of action of medication treatment for ADHD, has led to the **dopamine hypothesis,** which postulates that disturbances in the dopamine system may be related to the onset of ADHD. Fluorodopa positron emission tomography scans also support the dopamine hypothesis through the identification of low levels of dopamine activity in adults

457: D. Shoulder

Explanation:

AGE	X-RAY DONE FOR SKELETAL AGE DETERMINATION
Newborn	Knee
3-9 months	Shoulder
1-13 years	Hand, with wrist joint
12-14 years	Elbow & Hip

458: B. Autosomal dominant inheritance in most cases

Explanation: Clinical features of primary ciliary dyskinesia

Clinical Manifestations of Primary Ciliary Dyskinesia	
Respiratory Tract	**Genitourinary Tract**
Lung	Male and female infertility
Neonatal respiratory distress	
Chronic cough	
Recurrent pneumonia	
Bronchiectasis	
Middle Ear	**Left-Right orientation defects**
Chronic otitis media	Situs inversus
Conductive hearing loss	Heterotaxy
	Congenital heart disease
Paranasal Sinuses	**Central Nervous Sustem**
Neonatal rhinitis	Hydrocepnalus
Chronic mucopurulent rhinitis	Retinitis pigmentosa
Chronic pansinusitis	
Nasal polyposis	

Most cases are autosomal recessively inherited.

459: B. Pentavalent vaccine, Measles, Hepatitis B

Explanation: Remember that BCG is not given after 1 year of age and rotavirus vaccination series is not started beyond 14 weeks of age as per IAP and as per NIS, it is not given beyond 1 year of age.

National immunisation schedule

For Infants				
BCG	At birth or as early as possible till one year or age	0.1ml (0.05ml until 1 month age)	Intra-dermal	Left upper arm
Hepatitis B - Birth dose	At birth or as early as possible within 24 hours	0.5ml	Intra-muscular	Antero-lateral side of mid-thigh

OPV-O	At birth or as early as possible within the first 15 days	2 drpos	Oral	Oral
OVO 1, 2 & 3	At 6 weeks, 10 weeks & 14 weeks (OPV can be given till 5 years of age)	2 drops	Oral	Oral
Pentavalent 1, 2 & 3	At 6 weeks, 10 weeks & 14 weeks (can be given till one years of age)	0.5ml	Intra-muscular	Antero-lateral side of mid-thigh
Rotavirus	At 6 weeks, 10 weeks & 14 weeks (can be given till one years of age)	5 drops	Oral	Oral
IPV	Two fractional dose at 6 and 14 weeks of age	0.1ml	Intra dermal two fractional dose	Intra-dermal: Right upper arm
Measles / MR 1st Dose	9 completed months-12 months. (can be given till 5 years of age)	0.5ml	Sub-cutaneous	Right upper arm
JE-1	9 completed months 12 months	0.5ml	Sub-cutaneous	left upper arm
Vitamin A (1st dose)	At 9 completed months with measles-Rubells	1ml (1lakh IU)	Oral	Oral
For Children				
DPT booster-1	16-24 months	0.5ml	Intra-muscular	Antero-lateral side of mid-thigh
Measles / MR 2nd dose	16-24 months	0.5ml	Sub-cutaneous	Right upper arm
OPV Booster	16-24 months	2 drops	Oral	Oral
JE-2	16-24 months	0.5ml	Sub-cutaneous	Left upper arm
Vitamin A (2nd to 9th dose)	16-18 months. Then one dose every 6 months up to the age of 5 years.	2ml (2 lakh IU)	Oral	Oral
DPT Booster-2	5-6 years	0.5ml	Intra-muscular	Upper arm
TT	10 years & 16 years	0.5ml	Intra-muscular	Upper arm

460: C. Medial umbilical ligament

Explanation: Medial umbilical ligament arises from umbilical artery and Ligamentum teres arises from umbilical vein (intra-abdominal part).

461: B. Filaggrin

Explanation: **Severely dry skin is a hallmark of AD.** This results from compromise of the epidermal barrier, which leads to excess transepidermal water loss, allergen penetration, and microbial colonization. **Filaggrin**, a structural protein in the epidermis, and its breakdown products are critical to skin barrier function. Genetic mutations in the filaggrin gene family have been identified in up to 50% of patients with severe AD. Cytokines found in allergic inflammation, such as IL-4, IL-13, IL-22, IL-25, and tumor necrosis factor, can also reduce filaggrin expression.

462: C. Epigastric hernia

Explanation: Components of Pentalogy of Cantrell

- Omphalocele
- Anterior diaphragmatic hernia
- Sternal cleft with or without ectopia cordis
- Diaphragmatic pericardium defects (no diaphragmatic pericardium)
- Intracardiac defect: ventricular septal defect, diverticulum of the left ventricle, Tetralogy of Fallot

463: C. Canavan's disease

Explanation: Canavan disease:

- Autosomal recessive
- Deficiency of enzyme N-aspertoacylase
- Triad of macrocephaly, hypotonia and head lag

464: B. Metoclopramide

Explanation: The facial defect including cleft lip are characteristic teratogenic effect of isotretinoin. The drug used for treating acne should be used cautiously in reproductive age group after ruling out pregnancy.

465: A. Menkes disease

Explanation: Kinky hair is the key word in here.

- Menke's disese (TRICHO-POLIO DYSTOPHY) is copper metabolism disorder where there's mutation in ATP7A gene mutation
- XLR inheritance is seen
- High copper uptake in intestine but low transport across plasma ---- which leads to LOW TOTAL BODY COPPER LEVEL
- Symptoms-- hypotonia, pili-torti-like hair, seizures, malnutrition.
- RX ---- supportive
- In <2 years ---> Copper-histidine complex given

466: C. Central incisors, lateral incisors, canines, first molars

Explanation:

- Growth of the midface and lower face in children occurs gradually. Loss of the deciduous (baby) teeth is a more dramatic sign of maturation, beginning at around 6 years of age, after eruption of the first molars.
- Replacement with adult teeth occurs at a rate of about 4 per year.
- The mandibular, then maxillary central incisors are replaced at the same time or just following the eruption of the first molars at around 6–7 years of age.
- After this, the lateral incisors erupt, at around 7–8 years of age. The adult canines replace the baby teeth at around 9–11 years of age, and next to erupt are the first premolars and then the second premolars.
- The second molars erupt at around 12 years of age but the third molars (wisdom teeth) do not erupt until 17–21 years of age.

467: C. Rett syndrome

Explanation:

- Rett syndrome is a neurodevelopmental disorder of the grey matter of the brain that almost exclusively affects females.
- The clinical features include small hands and feet and a deceleration of the rate of head growth (including microcephaly in some).
- Repetitive hand movements, such as wringing and/or repeatedly putting hands into the mouth, are also noted. People with Rett syndrome are prone to gastrointestinal disorders and up to 80% have seizures.
- They typically have no verbal skills, and about 50% of individuals affected are not ambulatory. Scoliosis, growth failure, and constipation are very common and can be problematic.

468: B. Contralateral glaucoma

Explanation: Sturge Weber Syndrome has a triad of

- Port wine stain (restricted to one side of the face: mostly involving area supplied by ophthalmic division of trigeminal nerve)
- Ipsilateral glaucoma
- Ipsilateral leptomeningeal angiomas
- Complications may include hemiplegia, intracranial calcification (Rail Road/Tram Track appearance), Seizures: Focal, Mental retardation

469: B. Topical corticosteroids

Explanation: According to Nelson 20th Ed, topical corticosteroids are the cornerstone of anti-inflammatory treatment for acute exacerbations of atopic dermatitis/eczema

470: D. Acute airway obstruction from aspirated material

Explanation: Acutely after an aspiration there is a mechanical obstruction with irritation, which can cause bronchospasm. Later a chemical pneumonia and possibly bacterial infection may supervene.

471: A. CNS and disseminated TB

Explanation: BCG vaccine has a documented protective effect against CNS tuberculosis, i.e. tubercular meningitis, as well as disseminated TB. It does not prevent primary pulmonary infection as well as reactivation of latent infection.

472: B. DHEAS

Explanation: Fetal adrenal cortex shows high steroid sulfokinase activity and thus produces large amounts of DHEA and DHEA-S, particularly during mid-gestation (12 to 22 weeks). Placenta itself utilises fetal DHEA and DHEA-S as precursors to produce a variety of estrogens.

473: C. Omphalocele

Explanation: Omphalocele is a midline defect of anterior abdominal wall, with herniation of abdominal viscera into the base of umbilical cord. It is associated with chromosomal abnormalities in 43% cases.

474: D. Indomethacin

Explanation: Traditionally, corticosteroids, both oral and topical have been commonly used to treat infantile hemangiomas due to their inhibitory action on angiogenesis, with good results but have a problem of significant side-effects. Of late, Propranolol has been used in infantile hemangioma therapies, with good results and relatively lesser side effects than corticosteroids. Other therapies like surgery, radiotherapy, laser therapy, embolization, imiquimod, vincristine, and interferon-α therapies have been used in some patients with infantile hemangiomas.

475: C. Sturge Weber syndrome

Explanation: Causes of microcephaly

Causes of Microcephaly
Isolated microcephaly
Autosomal recessive, autosomal dominant or X-linked
Syndromic
Trisomies 21, 18, 13 Monosomy 1p36 deletion Syndromes: William, Cri-du-chat,Seckel, Smith Lemli Opitz, Cornelia de Lange, Rubinstein Taybi, Cockayne, Angelman
Structural diseases
Neural tube defect (anencephlay, hydranencephaly, encephalocele, holoprosencephaly) Lissencephaly, schizencephaly, polymicrogyria, pachygyria (macrogyria)
Metabolic disorders
Phenylketonuria, methylmalonic aciduria, citrullinemia Neuronal ceroid lipofuscunosis Maternal: diabetes mellitus, untrated phenylketonuria
Infections
Congenital: Cytomegalovirus, herpes simplex virus, rubella, varicella, toxoplasmosis, HIV, syphilis, enterovirus Meningitis
Teratogens

Alcohol, tobacco, marijuana, cocaine, heroin, toluene Antineoplastic agents, antiepileptic agents Radiology
Perinatal insult
Hypoxic ischemic encephalopathy, hypoglcemia
Endocrine
Hypothyroidism, hypopituitarism, adrenal insufficiency

476: B. Cerebral palsy

Explanation: Levine (POSTER) criteria for the diagnosis of cerebral palsy include

- P: Posturing
- O: Oropharyngeal problems
- S: Strabismus
- T: Tone abnormalities
- E: Evolutional maldevelopment e.g. Persistent primitive reflexes
- R: Reflexes are increased or brisk or exaggerated e.g. DTRs

Presence of any 4 out of 6 strongly suggests the diagnosis of cerebral palsy.

477: C. Inhalational route

Explanation: Most common route of spread to neonate of TB bacilli is by inhalation of infected secretions. Remember that hematogenous spread and aspiration of infected fluid are modes of acquiring congenital TB, not neonatal TB.

478: D. Bradycardia is seen

Explanation: Breath holding spells are brief periods of spells, where child has severe prolonged crying, followed by holding the breath in expiration. This is of two types: Pallid spell and cyanotic type. They are common between 6 moths to 3 years of age.

Pallid breath holding spell	Cyanotic breath holding spell
• Child turns pale and may become unconscious • Triggered by sudden fear, minor injury or painful stimulus • Less common • Due to parasympathetic overactivity • Bradyardia with asystole may occur in these • Treatment is conservative; atropine in severe cases	• Child turns blue and may become unconscious • Triggered by anger, vigorous crying or frustration • More common • Due to sympathetic overactivity • Tachycardia or normal heart rate • Treatment is conservative

479: D. Typhoid conjugate booster

Explanation: The question is NOT asking which vaccines can be given at 2 years of age as per IAP schedule. The question is asking which is recommended at 2 years. Since interpretation of question can vary from individual to individual and the language of question is a bit ambiguous, we shall stick strictly to IAP schedule which shows Typhoid conjugate booster at 2 years of age.

IAP immunization schedule for 2016

Age	Vaccines	Comments
Birth	BCG; OPV-0; Hep B-1	Administer these vaccines to all newborns before hospital discharge
6 weeks	DTwP 1; IPV 1; Hep-B 2; Hib 1; Rotavirus 1; PCV 1	DTP: • DTaP vaccine/combinations should preferably be avoided for the primary series • DTaP vaccine/combinations should be preferred in certain specific circumstances/conditions only • No need of repeating/giving additional doses of whole cell pertussis (wP) vaccine to a child who has earlier completed their primary schedule with acellular pertussis (aP) vaccine-containing products • Polio • All doses of IPV may be replaced with OPV if administration of the former is unfeasible • Additional doses of OPV on all supplementary immunization activities (SIAs) • Two doses of IPV instead of 3 for primary series if started at 8 weeks, and 8 weeks interval between the doses • Rotavirus: • 2 doses of RV1 and 3 doses of RV5 & RV 116E · • RV1 should be employed in 10 & 14 week schedule
10 weeks	DTwP 2; IPV 2; Hib 2; Rotavirus 2; PCV 2	Rotavirus: If RV1 is chosen, the first dose should be given at 10 weeks
14 weeks	DTwP 3; IPV 3; HiB 3; Rotavirus 3; PCV 3	Rotavirus: Only 2 doses of RV1 are recommended. If RV1 is chosen, the 2nd dose should be given at 14 week
6 months	OPV 1 Hep B 3	Hepatitis-B: The final (3rd or 4th) dose in the HepB vaccine series should be administered no earlier than age 24 weeks and at least 16 weeks after the first dose.
9 months	OPV-2 MMR-1	MMR: Measles-containing vaccine ideally should not be administered before completing 270 days or 9 months of life; The 2nd dose must follow in 2nd year of life; No need to give stand-alone measles vaccine

9-12 months	Typhoid conjugate vaccine	An interval of at least 4 weeks with the MMR vaccine should be maintained while administering this vaccine
12 months	Hep A 1	Hepatitis A: Single dose for live attenuated H2-strain Hep-A vaccine · Two doses for all inactivated Hep-A vaccines are recommended
15 months	MMR 2 Varicella 1 PCV booster	MMR: · The 2nd dose must follow in 2nd year of life · However, it can be given at anytime 4-8 weeks after the 1st dose Varicella: The risk of breakthrough varicella is lower if given 15 months onwards
16 to 18 months	DTwP Booster 1/ DTaP Booster 1; IPV Booster 1; Hib Booster 1	DTP: 1st & 2nd boosters should preferably be of DTwP
18 months	Hep A 2	Hepatitis A: 2nd dose for inactivated vaccines only
2 years	Booster of typhoid conjugate vaccine	• A booster dose of Typhoid conjugate vaccine (TCV), if primary dose is given at 9-12 months • A dose of Typhoid Vi-polysaccharide (Vi-PS) vaccine can be given if conjugate vaccine is not available or feasible; • Revaccination every 3 years with Vi-polysaccharide vaccine • Typhoid conjugate vaccine should be preferred over Vi- PS vaccine
4 to 6 years	DTwP Booster2/DTaP Booster 2; OPV 3; Varicella 2; MMR 3	Varicella: the 2nd dose can be given at anytime 3 months after the 1st dose. MMR: the 3rd dose is recommended at 4-6 years of age
10 to 12 years	Tdap/Td HPV	Tdap: is preferred to Td followed by Td every 10 years HPV: • Only 2 doses of either of the two HPV vaccines for adolescent/preadolescent girls aged 9-14 years; • For girls 15 years and older, and immunocompromised individuals 3 doses are recommended

480: A. Oral desmopressin

Explanation: **Pharmacologic therapy** is intended to treat the symptom of enuresis and thus is regarded as second line and is not curative. One form of treatment is **desmopressin acetate**, a synthetic analogue of antidiuretic hormone that reduces urine production overnight. This medication is FDA-approved and is available as a tablet, with a dosage of 0.2-0.6 mg at bedtime. In the past a nasal spray was used, but some children experienced hyponatremia and convulsions with this formulation and the nasal spray is no longer recommended for nocturnal enuresis.

481: B. Defect in beta- oxidation

Defect is in alpha oxidation,not beta oxidation. Others are correct.

482: B. Can be given to hemodynamically unstable babies

483: B. May be associated with Lead poisoning

484: D. Azithromycin is a good alternative

485: C. Pulmonary hypoplasia

Explanation: Bilateral renal agenesis is incompatible with extrauterine life and produces the Potter syndrome. Death occurs shortly after birth from pulmonary hypoplasia.

486: A. Mechanical ventilation for 48 hours

Explanation: **Indications for auditory screening (ABR or OAE) in children:**

1. Family H/O SNHL
2. In utero TORCH infections/congenital syphilis
3. Bacterial meningitis (particularly Pneumococcal & H. influenza)
4. H/o exchange transfusion for neonatal hyperbilirubinemia
5. Prolonged use of ototoxic drugs like Aminoglycosides, Loop diuretics
6. Apgar score 0 to 4 at 1 min & 0 to 6 at 5 min of life
7. Mechanical ventilation for >72 hrs
8. Suspected syndromic baby
9. Extreme prematurity

487: A. Down syndrome

Explanation:

- Most common genetic cause of mental retardation: Down syndrome
- Most common inherited cause of mental retardation: Fragile-X Syndrome
- Most common preventable cause of mental retardation: Congenital hypothyroidism

488: D. Black hairy tongue: Hemolytic anemia

Explanation: Abnormal tongue appearances and corresponding diseases

- Pale tongue: Anemia
- Strawberry tongue: Kawasaki disease, Scarlet fever
- Blue tongue: Cyanotic conditions, methemoglobinemia
- Yellow tongue: Jaundice
- Red, swollen, beefy tongue: Pellagra, Vit B12 def
- Smooth red tongue: Pernicious anemia
- Black hairy tongue: Fungal infections

489: A. Autosomal dominant

Explanation: All patients with typical achondroplasia have mutations at FGFR3 codon 380. The mutation maps to the transmembrane domain of the receptor and is thought to stabilize receptor dimers that enhance receptor signals, the consequences of which inhibit linear bone growth. Achondroplasia behaves as an autosomal dominant trait; most cases arise from a new mutation to normal parents.

490: C. Raised serum bilirubin

Explanation: Laboratory features of Reye syndrome

- Raised SGOT, SGPT, CPK, LDH, S.NH_3 (> 150 ug/dl)
- Raised Serum glutamate dehydrogenase
- Hypoglycemia, hypoprothrombinemia
- Normal serum bilirubin

491: D. 15q

Explanation: MFS is associated with abnormal production, matrix deposition and/or stability of fibrillin-1, a 350-kDa ECM protein that is the major constituent of microfibrils, with prominent disruption of microfibrils and elastic fibers in diseased tissues. The **fibrillin-1 (FBN1)** locus resides on the long arm of chromosome 15 (15q21), and the gene is composed of 65 exons.

492: C. Dyslexia

Explanation: Dyslexia may be the most common neurobehavioral disorder affecting children, with prevalence rates ranging from 5-10% in clinic- and school-identified samples to 17.5% in unselected population-based samples in the United States and other countries.

493: A. Biochemical evidence of acute liver injury lasting 12 weeks or less

Explanation: The currently accepted definition of fulminant hepatic failure (FHF) in children includes biochemical evidence of acute liver injury (usually <8 wk duration); no evidence of chronic liver disease; and hepatic-based coagulopathy defined as a prothrombin time (PT) >15 sec or international normalized ratio (INR) >1.5 not corrected by vitamin K in the presence of clinical hepatic encephalopathy, or a PT >20 sec or INR >2 regardless of the presence of clinical hepatic encephalopathy.

494: B. GRACILE syndrome

Explanation: The acronym GRACILE sums the most important clinical features, namely fetal growth restriction (birth weight about −4 SD), aminoaciduria (caused by Fanconi-type tubulopathy), cholestasis (with steatosis and cirrhosis), iron overload, severe lactic acidosis, and early death. The syndrome is associated with mutations of the complex III assembly factor BCS1L. The liver histology shows microvesicular steatosis and cholestasis with abundant iron accumulation in hepatocytes and Kupffer cells.

495: B. Staphylococcus aureus

Explanation: Altered cutaneous flora, with increased colonization by Staphylococcus aureus has been demonstrated. The increased staphylococcal carriage produces superinfection of skin lesions in AD. Colonization with methicillin-resistant strains of S. aureus is also more common than methicillin-sensitive ones in these patients.

496: D. Streptococcus mutans

Explanation: The group of microorganisms, mutans streptococci, is associated with the development of dental caries. These bacteria have the ability to adhere to enamel, produce abundant acid, and survive at low pH. Once the enamel surface cavitates, other oral bacteria (lactobacilli) can colonize the tooth, produce acid, and foster further tooth demineralization.

497: C. 5 years age

Explanation: Enuresis is defined as the repeated voiding of urine into clothes or bed at least twice a week for at least 3 consecutive months in a child who is at least 5 years of age and has a high prevalence in school aged children.

498: D. 48 months

Explanation:

- Even during the neonatal period, up to 10 of an infant's average of 16 hours of sleep per day are concentrated at night.
- This is reflective of an adaptation process which began in the third trimester of pregnancy.
- Daytime sleep progressively declines in infancy; the 12-month-old typically takes two 1-hour naps during the day.
- By the age of 3, most toddlers sleep 1 hour during the day, and the majority of 4-year-olds do not take a daytime nap.

499: D. Eagle-Barrett syndrome

Explanation:

- The constellation of cryptorchidism, posterior urethral valves, and abnormal abdominal musculature is called Eagle-Barrett syndrome.
- Another name is prune belly syndrome. The greatest morbidity comes from the poor amniotic fluid production, due to bladder outlet obstruction, with a resulting pulmonary hypoplasia.
- Cushing's triad are hypertension, bradycardia, and widened pulse pressure. This is seen as terminal findings associated with increased intracranial pressure.
- VATER association has multiple anomalies, none of which are the three mentioned. VATER is a mnemonic which stands for Vertebral anomalies, Anal atresia (imperforate), Tracheo-esophageal fistula, and Renal anomalies (the R also indicated Radial anomalies).

500: A. Vernal keratoconjunctivitis

Explanation: Allergen immunotherapy is **contraindicated** in children undergoing β-blocker therapy as well as those with certain immunologic or autoimmune disorders, allergic bronchopulmonary aspergillosis, hypersensitivity pneumonitis, severe psychiatric disturbance, or a medical condition that would impair the ability to survive an allergic reaction.